

**A qualitative study exploring the specific everyday
social and environmental challenges faced by
community dwelling people living with posterior
cortical atrophy (PCA)**

“They don’t understand”

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A qualitative study exploring the specific everyday social and environmental challenges faced by
community dwelling people living with posterior cortical atrophy (PCA)

DECLARATION STATEMENT

I, Pamela Turpin, confirm that the work presented in this thesis is my own. Where information has been derived from other sources, I confirm that this has been indicated in the thesis.

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Abstract

Posterior Cortical Atrophy (PCA) is a rare progressive degenerative neurological condition where individuals experience serious vision and perception difficulties. In most cases, the underlying cause for these problems is Alzheimer's disease. Unlike typical Alzheimer's, the damage to brain cells is particularly focused at the back of the brain and in the early stages of the condition, day to day memory remains intact. Diagnosis of PCA is often protracted and to date, the specific needs of such individuals have tended to be overlooked.

The aim of the study is to contribute to the knowledge base around visual impairment caused by PCA. This study recognised the need for more qualitative inquiry into the specific challenges faced by individuals with this condition. Understanding effective types of improvements to a person's social and physical environment may improve the wellbeing of community dwelling people experiencing this debilitating condition.

To explore practical 'real-life' problems and issues, a qualitative research study using in-depth interviews with four community dwelling individuals living with PCA and their caregivers is undertaken. A person-centred philosophy is emphasised to enable the voices of these individuals to be heard. Phenomenological analysis is used to gain a better understanding of the subjective experiences of living with PCA and/or caring for somebody with this condition.

The key message from participants is poor understanding of PCA by health and social professionals as well as the general public, inhibiting the provision of information and support for the people with PCA and their caregivers.

Finally, the results of this investigation have been used to generate the basic tenets of a best practice guide for supporting people living with PCA and their caregivers.

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Keywords: posterior cortical atrophy; dementia; Alzheimer's disease; vision loss; perception; physical environments; case study design; qualitative research; care model.

Word Count: 292.

Impact Statement

Posterior Cortical Atrophy (PCA) is a rare progressive degenerative neurological condition where individuals experience serious vision and perception difficulties. The symptoms of PCA usually appear in people in their late 50s and early 60s. In most cases, the underlying cause for these problems is Alzheimer's disease. It has been estimated that 5% of patients seen in specialty dementia diagnostic centres have PCA and up to 13% of individuals presenting with early onset AD have the syndrome. With cases of AD predicted to substantially increase over the next 30 years, it is reasonable to presume cases of PCA will also rise proportionately.

PCA presents in a very different way from typical AD with individuals retaining memory and insight well into the later stages of the condition. Although biomedical studies have reported on **WHY** visual symptoms are occurring, there is a paucity of studies describing **HOW** these symptoms impact on the individual living with PCA at an emotional, physical and personal level.

The aim of this qualitative research study has been to contribute to the knowledge base around visual impairment caused by PCA. By using in-depth interviews with individuals living with PCA and their caregivers, a better understanding of the subjective experiences of those living with this condition has been achieved.

Benefits inside academia

The benefit of this research has been to contribute to the knowledge base around visual impairment caused by PCA. There is currently limited understanding of the subjective experiences of people with PCA and their caregivers. Larger studies of this population would further inform researchers and policy makers about the specific needs of this population. This study has highlighted areas needed for future research particularly wayfinding around the home, developing assistive technologies to enable people with PCA to continue to undertake activities of daily living, and developing a physical exercise programme to enhance their wellbeing.

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Such research would not only benefit people with PCA but also allow those who have both visual and cognitive impairments to remain as independent.

Benefits outside academia

The beneficial impact of this study could occur locally, regionally and nationally or internationally with the use of the proposed best practice guide specifically for PCA. By engaging with public policy makers, service delivery practitioners as well as people with PCA, the guide could be refined and attached to national dementia strategies. This would enable information, training and support to be mainstreamed for people with PCA and their caregivers to access. It would also increase awareness of this condition to health and social care professionals and improve care practices.

Disseminating outputs

Awareness of the day to day challenges of living with PCA could be disseminated through scholarly journals as well as specialist and mainstream publications, presentations at national and international conferences, and teaching seminars for health and social care professionals as well as caregivers.

467 words

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Dedication

This study is dedicated to anyone who has been touched by the experience of Posterior Cortical Atrophy as well as the myriad of researchers who are now investigating this challenging condition and bringing it to the world's attention.

Acknowledgement

I would like to thank all the participants and their caregivers who welcomed me into their homes and shared their challenges of living every day with posterior cortical atrophy. Without the honest retelling of their experiences, this thesis could never have been achieved.

I would like to thank Professor Nick Tyler for all his inspirational teaching, advising and support.

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Entry vignettes

“She looks for things like a blind person. That’s what she does. She’s looking but not seeing.” (Caregiver of person with PCA)

“I still sometimes in the morning get up and think ‘oh I’ll do such and such’ and then I either start or think ‘I can’t do it, I know I can’t do it. I’ll make a mess of it’. So, my confidence is way down. I’ve lost it. I don’t trust myself.” (Person living with PCA)

“...there is a sense of isolation due to the rareness of the condition and the fact is it gets a little difficult to bear if your recourse is to your doctor or a psychologist or counselling because in most of cases they don’t understand PCA.” (Person living with PCA)

“I am very, very sad, very sad. It’s a mixture of anger at the situation, anger with myself because I can’t handle it. Great sadness. In fact, a dreadful sadness. There has been no absence of tears.” (Caregiver of person with PCA)

“I was upset because right from the beginning they kept saying ‘there’s nothing we can do. There’s nothing we can do’. Everybody says the same.” (Person living with PCA)

1.0 Chapter One: Setting the scene

This chapter introduces my motivation for undertaking the study. An outline of the investigation is given. The structure of the thesis is outlined to assist the reader in the navigation of the document. Finally, a table of abbreviations and acronyms is displayed.

1.1 Introduction

I have worked with people living with dementia both at a personal and professional level for over twenty years and am passionate about trying to maintain a good quality of life for these individuals throughout their dementia journey. During my experiences of supporting care home residents living with dementia, I noticed that many had age related visual difficulties as well as cognitive decline, but these additional problems were often not fully considered by care staff due to lack of awareness training regarding their specific needs. This lack of understanding around age related sensory challenges of people living with dementia prompted a MA research study which focused on the needs of people with sensory and cognitive morbidities. The results from the investigation enabled me to assist in the development of a training programme for formal and informal carers about visual and cognitive losses that occur concurrently, as well as the production of a low vision care passport for use by residents in an assisted care facility.

During my studies, an additional group of individuals came to my attention who struggled with visual and perceptual problems due to dementia itself, rather than an eye disease occurring alongside cognitive decline. However, because there was little ongoing research into this phenomenon at that time, I subsequently focused my research only on age related eye diseases that can occur in conjunction with dementia.

When I was given the opportunity to undertake a PhD at UCL I decided I wanted to take the study of vision loss with dementia further. Whilst attending a meeting at the Dementia Research Centre, UCL Institute of Neurology in London, I was able to talk with people, many in their fifties or early sixties, who were experiencing a rare form of Alzheimer's disease

(AD) known as posterior cortical atrophy (PCA). This was the condition I had first come across in the 1990s and was now being studied more widely by medical researchers. These participants eloquently described the visual and perceptual problems they were experiencing including difficulties with reading; recognising objects or faces; problems with spatial awareness resulting in difficulty descending stairs, driving or trying to pick something up; double vision; increased sensitivity to bright lights or shiny surfaces or struggling to see clearly in low or fading light levels (Levine, Lee, & Fisher, 1993; Bokde, Pietrini, Ibanez, & *et al*, 2001; Chan, Crutch, & Warrington, 2001; Zakzanis & Boulos, 2001; Shakespeare, Yong, Foxe, Hodges, & Crutch, 2015; Crutch, Walton, Carton, & Shakespeare, 2017) .

I learned that the well-known author, Terry Pratchett, had also lived with PCA from 2007 to 2015. He called it his '*embuggerance*' describing it thus:

"I have the opposite to a superpower; sometimes I cannot see what is there. I see the teacup with my eyes, but my brain refuses to send me the teacup message. ... First there is no teacup and then, because I know there is a teacup, the teacup will appear the next time I look." (Pratchett: 2015 pp31/32).

From discussions with researchers at this meeting, it appeared that although clinical research into this condition was now being undertaken worldwide, there was a paucity of investigations regarding the subjective experiences of people living with PCA. I determined to explore if a study examining the specific everyday social and environmental challenges faced by people living with PCA might be a useful addition to the literature.

1.2 Outline of the research

The thesis is structured as shown in **Table One**:

Table 1: Organisation of the thesis

Section	Detail	Page
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The aim of the study is to contribute to the knowledge base around visual and perception challenges caused by PCA. The investigation focuses on a small number of couples living with PCA in the community, one living with

the disease and one being their caregiver. The research is being undertaken to improve understanding of the subjective experience of living with PCA and determine how medical and social care can be tailored to improve the support available to this populace.

1.3 Abbreviations

Table Two shows abbreviations that appear in this document.

Table 2: Abbreviations

ADLs	Activities of daily living is a term used in healthcare to refer to people's daily self care activities.
AT	Assistive technology (AT) is assistive, adaptive, and rehabilitative devices for people with disabilities or the elderly population. People who have disabilities often have difficulty performing activities of daily living (ADLs) independently, or even with assistance.
DLB	Dementia with Lewy bodies. Lewy bodies are small spherical protein deposits and are also found in the basal ganglia neurones in brains of people with Parkinson's disease affecting movement.
Dry AMD	Dry age related macular degeneration occurs when drusen accumulates in the macula at the back of the eye causing central vision loss. The presence of many drusen is the early sign of dry age-related macular degeneration, the most common form of the condition.
EOAD	<i>early-onset</i> Alzheimer's Disease if diagnosis occurs before the age of 65 years.
FTL	Fronto-temporal dementia covers a range of conditions (including Pick's disease) caused by degeneration of the frontal and temporal lobes. Clinical characteristics include changes in behaviour, mood and personality with relative preservation of memory and cognitive functions.
LOAD	Late onset Alzheimer's disease if the diagnosis occurs after the age of 65 years.
MCI	Mild cognitive impairment means minor problems with mental abilities, such as memory or thinking. MCI is not a type of dementia.

MD	Mixed dementia occurs when two subtypes exist together (for example AD and VD).
NDS	National Dementia Strategy provides a strategic framework within which local health and social care services can deliver quality improvements to dementia services.
PCA	Posterior Cortical Atrophy is a rare atypical dementia that presents in people in their 50s and early 60s. It is characterised by a striking decline in higher order visual-processing abilities and other cognitive impairments due to progressive atrophy in the parietal, occipital and occipitotemporal regions of the brain.
tAD	Typical Alzheimer’s Disease has a distinct pattern of amyloid plaques and neurofibrillary tangles which cause progressive neurodegeneration usually starting with the hippocampus and entorhinal cortex, progressing to the neocortex of the brain, particularly the temporal, frontal and parietal regions impacting on their associated functions.
UCL	University College London.
UK	United Kingdom.
VaD	Vascular Dementia also known as multi-infarct dementia. It is caused by insufficient blood supply to the brain resulting in the death of brain cells.
Wet AMD	Wet age related macular degeneration occurs when blood vessels grow abnormally under the retina which may leak and cause scarring on the macular.

The next chapter aims to give a brief general explanation about dementia and age related visual impairment in order to compare their characteristics with that of posterior cortical atrophy (PCA) as well as problems that arise when a person has vision loss alongside cognitive decline due to either age related eye disease or because of dementia itself. I will then provide the reader with detailed information about posterior cortical atrophy (PCA) and justify why I have decided to undertake research to explore its effects on the daily lives of both people with the condition, and their family caregivers.

2.0 Chapter Two: Background to the study

2.1 Introduction

This chapter aims to provide the reader with detailed information about posterior cortical atrophy (PCA) and to justify why I have decided to undertake research to explore its effects on the daily lives of both people with the condition, and their family caregivers. To address this issue, I will investigate current academic understanding regarding the nature of this rare dementia variant. This includes seeking to understand in what ways PCA differs in presentation from more common forms of dementia; identify the characteristics of its physical manifestations; and determine what knowledge exists regarding how living with progressive visual and perceptual difficulties impacts on individuals and their caregivers.

First I will set out the research protocol I have used to locate and explore current thinking around PCA. A brief general explanation about dementia, and age related visual impairment is given in order to compare their characteristics with that of PCA. Next, problems that arise when a person has vision loss alongside cognitive decline due to either age related eye disease or because of dementia itself are identified. An in-depth exploration is then presented regarding current medical and clinical knowledge of PCA and the differences between the symptoms tAD and PCA are compared. Alzheimer's disease has been chosen for this comparison because present medical studies have determined that PCA is more likely to have underlying AD pathology (See [section 2.6](#)) Finally gaps within academic research that I believe deserve more research attention are suggested.

2.2 Gathering the literature

To locate research papers I used a search strategy to identify published academic studies published in peer reviewed journals as well as 'grey' literature. (Grey literature includes web pages, articles, reports, fact sheets

and theses, which are not published by commercial or academic publishers).

For published literature UCL Explore database was searched.

On-line research registers included:

- AMED (Allied and Complementary Medicine)
- BMJ
- CIMAHL Plus
- Cochrane Library
- EMBASE
- Pubmed
- SCOPUS

The internet, conference proceedings and professional journals were also explored.

The search was limited to national and international literature published in English over the last fifty years.

Keywords used in database searches included: dementia; posterior cortical atrophy; Alzheimer's disease; age related eye disease; sight loss with dementia; dementia care practice; subjective experiences of dementia.

198 articles focusing specifically on PCA were identified. These were scanned for relevancy and duplication. This enabled a reduced number of 103 texts which I have used to inform the background material. It was interesting to note that the majority of papers found were medical or clinical research studies. Six articles explored the non-pharmaceutical management of PCA and two of these focused on the subjective experiences of people with PCA and/or their caregivers.

The next section gives an overview of dementia, outlines the condition, the most common presentations.

2.3 Dementia

The rise of population ageing is expected to lead to dramatic increases in the prevalence of dementia which will affect all regions of the world (Norton, Matthews, & Brayne, 2013; Livingston, *et al.*, 2017). In the UK, an

estimated 800,000 people are currently living with dementia (Parkin & Baker, 2016) and these numbers are predicted to double by 2040 (Department of Health (a), 2015). It has been reported that one in every three UK children born in 2015 will develop a dementia during their lifetime (Alzheimer's Research UK, 2015). Two thirds of people with dementia live in the community while one third live in residential care (Department of Health, 2013; SCIE, 2015).

Dementia as a concept has evolved over more than 2,000 years from the notion of inevitable age-related cognitive decline (Scafer, 2002) to a modern scientific understanding of its clinical and pathologic features (Grand & Feldman, 2007).

Dementia is an umbrella term used to describe many different conditions that cause a collection of symptoms, of which the most common include: memory loss, confusion, mood and personality changes, problems with planning, orientation and reasoning skills as well as the gradual inability to perform daily living tasks (Moore, 2009; Ray & Davidson, 2014). Currently, in most cases, it is an incurable disease with an extremely variable time course (Rahman, 2014).

Over 200 subtypes of dementia have been identified (Stephen & Brayne, 2008) but the most common are Alzheimer's Disease (AD), vascular dementia (VD), dementia with Lewy bodies (DLB), frontotemporal dementia (FT) and mixed dementia (MD). MD occurs when two subtypes exist together (for example AD and VD) (Dementia UK, 2018).

Development of brain scanning techniques have allowed specialists to improve comprehension of the structural and functional changes that occur within the brain and assist in the differential diagnosis types of the disease (Jolley, 2010).

The introduction of the UK national dementia strategy in 2009 has led to a greater emphasis on earlier diagnosis, support and treatment for people with dementia and their family and caregivers (Department of Health, 2009) but the time it takes to make a diagnosis can vary tremendously. In the early stages of the disease, an individual may be monitored for up to 12

months before a diagnosis can be made, and if scans and other diagnostic tests are required, the Alzheimer's Society has estimated that this can take between 4 to 12 weeks, depending on waiting lists, before a diagnosis can be confirmed (Alzheimer's Society, 2017).

A clear benefit to the person with dementia on receiving an early diagnosis is that they are able to access support services, develop a care plan, organise power of attorney and end of life care wishes while they are still able to be in control of these procedures (Stanley & Cantley, 2001)

2.3.1 Alzheimer's Disease

At a lecture in November 1906, the German neurologist Alois Alzheimer described a form of dementia affecting a 51 year old woman called Auguste D who was experiencing memory and language impairments and behavioural changes. At the suggestion of his colleague, Emil Kraepelin, a renown German psychiatrist at the time, this disorder became known as Alzheimer's disease (Maurer, Volk, & Gerbaldo, 1997). Today, it is the most prevalent form of dementia accounting for two thirds of dementia cases in the UK (Alzheimer's Research UK, 2018).

AD is classed as *early-onset* (EOAD) if diagnosis occurs before the age of 65 years, and *late-onset* (LOAD) if diagnosis occurs in a person over 65 years.

Late onset AD (LOAD), alone or in combination with other illnesses, accounts for between 60 per cent and 80 per cent of all cases (Alzheimer's Association, 2017).

An estimated 42,000 younger people under the age of 65 years are living with dementia in the UK (Alzheimer's Society, 2015). Alzheimer's is the most common type of dementia in younger people and early onset AD (EOAD) affects around a third of younger people with dementia (Mendez, 2019). It has been found that younger individuals are more likely to have an *atypical* form of AD than older people. Problems with vision, speech or behaviour may be the first presenting symptoms as opposed to memory loss. It has been estimated that such atypical forms of AD account for up

to one-third of all AD cases in younger people in contrast with 5% of individuals with LOAD (Alzheimers Society, 2015).

Draper et al (2016) examined factors determined the time taken to diagnosis people with EOAD by mapping their diagnosis pathways. Out of 88 individuals tracked, researchers found that individuals had a significantly longer time from onset of symptoms to their first consultation, median time being 2.3 years. It also revealed that the dementia diagnosis was appreciably longer if a participant presented with mild cognitive impairment (MCI) or depression. When the dementia was not AD or FTD, final diagnosis of the type of dementia could take 4.7 years.

Less than five per cent of AD cases are caused by an inherited familial disorder where there is a mutation of three genes. Such instances occur more often in early-onset cases (Hughes, 2011).

Typical Alzheimer's Disease (tAD) has a distinct pattern of amyloid plaques and neurofibrillary tangles which cause progressive neurodegeneration usually starting with the hippocampus and entorhinal cortex, progressing to the neocortex of the brain, particularly the temporal, frontal and parietal regions impacting on their associated functions. Usually the primary motor, sensory and visual cortex are spared until late in the disease (Quadi, Alipour, & Beattie, 2007; Stephen & Brayne, 2008).

The condition is characterised by a steady and progressive decline in memory and cognitive functions including impaired visuospatial skills, language deterioration, poor judgement, recognition difficulties and personality changes. In the most advanced phase, the person becomes totally dependent on others for their care and support (Hughes, 2006; Adams, 2008).

Although no accurate prediction of length or severity of illness progression can be made for any individual, studies indicate that people age 65 and older survive on average of 4 to 8 years after a diagnosis of Alzheimer's dementia, yet some live as long as 20 years reflecting Alzheimer's slow but steady progression of decline (Larson, Shadlen, Wang, McCormick, *et al.*,

2004; Ganguli, *et al.*, 2005; Broadaty, Seeher, & Gibson, 2012; Tom, *et al.*, 2015).

2.3.2 Vascular dementia

The next most common type is vascular dementia (VaD), also known as multi-infarct dementia. This condition affects around 150,000 people in the UK (Alzheimer's Society, 2014). It is caused by insufficient blood supply to the brain resulting in the death of brain cells. It can follow a large stroke caused when a blood vessel becomes blocked (ischaemic stroke), or a bleed from a blood vessel within the brain (haemorrhagic stroke). It can also be caused by a series of mini strokes (multi-infarct stroke) which a person may not even know is happening (Adams, 2008; Hughes, 2011).

The main characteristics of VaD are loss of short-term memory, poor concentration and communication. The individual may also display physical symptoms such as limb weakness or paralysis. Personality is relatively preserved but there is an increased likelihood of unpredictable behaviour and changeable emotions. For instance, people with VaD have a greater awareness of their condition than people with tAD which can lead to depressive moods (Alzheimer's Knowledge Exchange, 2016). Onset is usually abrupt, characterised by instances of sharp decline alternating with periods of stability. Life expectancy is poor (Stephen & Brayne, 2008).

2.3.3 Dementia with Lewy bodies

Dementia with Lewy bodies (DLB) accounts for one in 25 cases of people living with dementia in the community (Vann Jones & O'Brien, 2014). Lewy bodies are small spherical protein deposits and are also found in the basal ganglia neurones in brains of people with Parkinson's disease affecting movement (Adams, 2008; The Lewy Body Society, 2015).

DLB is a progressive condition, developing over several years. People with DLB experience vivid visual hallucinations, spatial disorientation, impaired recent memory with fluctuations in mental performance and communication difficulties. Individuals exhibit Parkinsonism symptoms

such as slow movements, muscle stiffness, trembling limbs, shuffling gait and a mask-like expression. They are also prone to falling (Jolley, 2010).

2.3.4 Fronto-temporal dementia

Fronto-temporal dementia (FTL) covers a range of conditions (including Pick's disease) caused by degeneration of the frontal and temporal lobes. Clinical characteristics include changes in behaviour, mood and personality with relative preservation of memory and cognitive functions (Jolley, 2010; Alzheimer's Society (b), 2016). Younger people under 65 years are more likely to be affected and a family history of occurrence is more probable than for other types of dementia (Warren, Rohrer, & Rossor, 2013). In the later stages of the disease, damage to the brain becomes more widespread, resulting in symptoms comparable with tAD (Adams, 2008).

2.3.5 Impact of being given a diagnosis of dementia

Because individual responses may be quite different, the manner in which the diagnosis is received is of paramount importance. Lecouturier, *et al.*, (2008) suggested that there is limited guidance on the most appropriate way to disclose a diagnosis of dementia. If given badly, there can be poor retention of information and lack of understanding.

The impact of receiving a dementia diagnosis has been described in a study by Pratt & Wilkinson (2001) who stressed the importance of considering the impact of both psychological and social factors on people's lives when giving the diagnosis. Books written by people with dementia also describe their experiences of diagnosis (McGowan, 1993; Rose, 1996; Bryden, 2005).

Pre diagnostic counselling can be beneficial resulting in gradual realisation of the probable causes of symptoms (Manthorpe, 2001). Relief may then be expressed when the final disclosure is given:

“I was not surprised to find out I was suffering from dementia. I had been forewarned about it, and had a definite feeling that that was probably what it was.” (Kabel, 2001) p570

The impact of a diagnosis of dementia will affect family and friends as well as the person that has been diagnosed. Although diagnosis can help the caregiver understand particular challenges, such as behaviours that are difficult to understand, dementia can put a huge strain on relationships both interpersonal and social (Derksen, Vernooij-Dassen, Gilliss, Olde-Rollert, & Scheltens, 2005).

The losses felt by the experience of dementia have been well documented. A meta-synthesis of qualitative dementia research found that living with dementia comprises of undergoing both negative and positive emotions (Górska,, Forsyth, & Maciver, 2018). The inner fear of losing oneself once you have a label of dementia can be exacerbated when you lose the role you once had either in your home, career, or in society (Davis, 1989; Holst & Hallberg, 2003; SCIE, 2015; Whitman, 2016). The inability to undertake tasks can cause frustration, anger and embarrassment (Caddell & Clare, 2011) although positive feelings such as love and optimism have also been expressed (Karlsson, Savenstedt, Axelsson, & Zingmark, 2014).

The worry of rejection can be foremost in people’s minds due to lack of public awareness and the stigma associated with a mental health illness. Lack of awareness about dementia can result in people and organisations not supporting the abilities that people with dementia retain.

Despite policy initiatives regarding improving the care of people with dementia (DH/SCLGCP/SCP/OP&D, 2010; Department of Health (b), 2015) and the UK National Dementia Strategy (Department of Health, 2009) there is a need for more educational programmes amongst the general public in order to raise awareness and understanding of dementia (Cahill, Pierce, Werner, Darley, & Bobersky, 2015).

2.3.6 Interim Summary

This section has given an overview of dementia, outlined the condition, the most common presentations and considered the emotional impact receiving a dementia diagnosis can make on individuals with dementia and their caregivers. **Table 3** outlines key points from section 2.3.

Key Points from Section 2.3

Table 3: Key points from Section 2.3

✓	800,000 people in the UK are currently living with dementia and these numbers are predicted to double by 2040.
✓	Dementia is an umbrella term describing progressive terminal conditions caused by deterioration of brain tissue.
✓	The most common causes of dementia are Alzheimer’s disease (AD), vascular dementia (VaD), dementia with Lewy bodies (LBD) and fronto-temporal dementia (FTD).
✓	Mixed dementia (MD) occurs when two subtypes exist together (for example AD and VD).
✓	Early onset AD (EOAD) affects around a third of younger people under 65 years.. It has been found that younger individuals are more likely to have an <i>atypical</i> form of AD than older people.
✓	The introduction of the UK national dementia strategy has led to a greater emphasis on earlier diagnosis, support and treatment for people with dementia and their family and caregivers. However, the time it takes to make a diagnosis can vary tremendously.
✓	There is a need for more educational programmes amongst the general public in order to raise awareness and understanding of dementia

The focus of this research study is posterior cortical atrophy (PCA) which often presents as an atypical form of AD. The study will be exploring the visual difficulties people with PCA experience, most of whom will be over 50 years of age. This means that age related visual changes may also be developing in this population. To help gain understanding in this area, I will now turn attention to age related eye conditions occurring alongside a

diagnosis of typical dementia. The more detailed consideration of PCA will then be taken up in [section 2.6](#).

2.4 Age-related eye diseases existing with dementia

2.4.1 Introduction

Until the 1980s, little attention was given to visual problems that may affect individuals with dementia. (Morse, 2000) advised that because the prevalence of both dementia and vision impairment increases with age, the degree of visual impairment among people with dementia may be greater than in the general population. (Trigg & Jones, 2005) estimated that 2 per cent of people over 75 years had both conditions cautioning that this figure was likely to be an underestimate because visual impairment studies would not have accounted for individuals considered '*untestable*'. Most eye testing requires verbal responses from the patient so would exclude people with dementia who may have lost their ability to communicate effectively or lack insight into what is being asked of them. It is likely that such individuals would stop receiving regular ophthalmologic examinations or using correctly prescribed spectacles or visual aids resulting in any sight problems being untreated due to being ignored or undetected (Koch, Datta, Makhdoom, & Grossberg, 2005).

Visual impairments make it harder for people with dementia to see properly, understand the environment they are in and possibly increase confusion. It will also limit their participation in everyday activities like watching television, reading, knitting, doing jigsaws, as well as resulting in them having a higher risk of falls and difficulties with doing everyday tasks such as walking, dressing, eating and drinking.

Visual impairment and blindness due to age-related eye disorders is common. Several eye studies have been conducted to determine the prevalence and causes of visual impairment in the developed world (Leibowitz, Krueger, Maunder, *et al.*, 1980; Mitchell, Hayes, & Wang, 1997; Munier, Gunning, Kenny, & O'Keefe, 1998; Buch, Vinding, & Neilsen, 2001; Evans, *et al.*, 2002; Minassian & Reidy, 2009). These studies either used registration figures for blind people or were based on a sample of

population. Significantly, the leading causes of vision loss reported in all studies were the same: cataract, macular degeneration, glaucoma and diabetes. (Tideiksaar, 2002/3) referred to these as the “*Big Four*” noting that these conditions *caused “a multitude of symptoms that increased fall risk”* (p22).

2.4.2 Cataract

Cataracts are caused by the lens of the eye becoming opaque preventing light reaching the retina resulting in poor vision. People with cataracts also experience increased sensitivity to glare and great difficulty in seeing at low light levels. This reduction in sight is gradual and sometimes accepted as just ‘*old age*’ and left untreated. Cataract surgery has a high level of success and has minimal complications (Minassian & Reidy, 2009). Evans, Fletcher, & Wormald (2004) estimated that 26 per cent of cases of sight loss in people aged over 75 years were due to cataracts which could be successfully treated in almost 90 per cent of all cases resulting in improved visual function, mood and visual quality of life (Ortiz-Figureoa, *et al.*, 2016).

AD and cataracts frequently co-exist (Kumar & Seet, 2016; RNIB, 2020). Studies have suggested that these degenerative processes may share common aetiological mechanisms (Jefferis, Mosimann, & Clarke, 2011; Fukuoka, 2015). For example, risk factors for cataracts, which include ageing, female gender, smoking, diabetes and obesity as well as lower socioeconomic class and associated lower educational attainment, are also risk factors for dementia (Livingston, *et al.*, 2017; Liu, *et al.*, 2020). Jefferis, Mosimann, & Clarke (2011) have suggested that because of such common risk factors, people living with dementia may also have a higher incidence rate of cataract highlighting that exploration of possible interactions between both conditions is a neglected area of research.

If cataract removal is not an option due to frailty, it is vital that caregivers are aware of the visual difficulties individuals may be experiencing. They could then consider making appropriate sensory adjustments to the social and physical environment to improve general wellbeing. This could mean

increasing light levels and bold colour contrast in toilets, corridors, furniture and crockery to help enable functional abilities.

2.4.3 Age Related Macular Degeneration (AMD)

It is estimated that more than half a million people in the UK are affected by AMD and this is predicted to double by 2050 (Macular Society, 2017). The huge rise in the ageing population is the main reason for the increase in the condition. One in 250 people has sight loss due to AMD at 60 years which quadruples to one in four people at the age of 90 (Owen, *et al.*, 2012; Macular Society, 2017). Changes occur in the macula at the back of the eye causing central vision loss (RNIB, 2016). Drusen, yellow deposits under the retina made up of lipids, is a fatty protein. The presence of many drusen is the early sign of dry age-related macular degeneration, the most common form of the condition. Less prevalent is wet AMD. This occurs when blood vessels grow abnormally under the retina which may leak and cause scarring on the macular (Porter, 2017).

Alzheimer's disease and AMD have been found to have common characteristics (Zetterberg, *et al.*, 2008). The presence of amyloid β has been found both in the plaques seen in the brains of dementia patients and the drusen of AMD patients (Ohno-Matsui, 2011). However, further research is required to determine if AMD is significantly more common in patients with AD as current findings are not conclusive (Keenan, Goldacre, & Goldacre, 2014; Tsai, Chen, Huang, Yaun, & Leu, 2015).

Loss of central vision reduces individuals' abilities to undertake activities of daily living as well as reading, watching television, recognising faces, eating independently and increases trips and falls risk.

2.4.4 Glaucoma

Glaucoma is the second leading cause of blindness in the world (Glaucoma Research Foundation, 2017). More than 500,000 people in England and Wales alone are affected by the condition (Association of British Dispensing Opticians, 2018) .

The condition is caused by increased aqueous humour due to drainage problems resulting in a build-up of pressure with the eye (IGA, 2012). The condition can be treated with daily eye drops or an artificial drainage hole created in the eye. It is important that glaucoma is detected early to prevent damage to the retina and optic nerve fibres causing peripheral visual field loss. Individuals can experience tunnel vision causing difficulties with mobility and identification of trip hazards (Glaucoma Research Foundation, 2016).

Links between AD and glaucoma have been found although genetic and clinical relationships remain unclear (Ward & Petersen, 2013; Meszaros, 2014). For example, some studies have suggested that there is an increased rate of glaucoma occurrence in individuals with Alzheimer's disease (Vickers, et al., 2002; Wostyn, Audenaert, & De Deyn, 2009) whilst others have contended that the condition is not associated with increased risk of developing AD (Ressiniotis, Griffiths, Birch, Keers, & Chinnery, 2004; Kessing, Lopez, Anderson, & Kessing, 2007).

The benefits of early detection of glaucoma in individuals with dementia have been highlighted (Pankow, Pryor, & Luchins, 2009). The course of sight loss due to glaucoma was found to be swifter and more aggressive in patients with dementia than in those without dementia possibly due to non-compliance regarding daily eye drop medication (Sinclair, Hinds, & Sanders, 2004; Valenti, 2004).

2.4.5 Diabetic Retinopathy

Diabetic retinopathy is the most common form of diabetic eye disease and can affect people who have had diabetes for several years. It occurs when changes in the blood glucose level cause retinal blood vessels to swell and leak blood and fluids into the rear of the eye. In other cases, abnormal blood vessels grow on the surface of the retina. If left untreated, an individual has an increased risk of blindness (National Eye Institute, 2015).

Amongst people with dementia, prevalence rates of diabetes are high. Study results vary from 6 per cent to 39 per cent (Bunn, *et al.*, 2014). Although the link is not yet fully understood, Type 2 diabetes has been

found to increase the risk of dementia (Feng-Ping, Kun-Pei, & Hsu-Ko, 2009; Livingston, *et al.*, 2017). People with dementia may be unable to recognise the signs of diabetes or to communicate their concerns effectively to their caregivers (Scrutton & Brancati, 2016).

Once diagnosed, individuals with diabetes need to maintain strict procedures regarding medication, diet and medical appointments to ensure that blood glucose levels remain as stable as possible. Caregivers play a vital role. However, they often encounter difficulties in ensuring that the person with dementia and diabetes complies with the treatments designed to manage their condition (McDonald, Hertz, Unger, & Lustik, 2009). Studies have recorded caregivers advising that they feel they must be “*on duty 24 hours a day*” (National Alliance for Caregiving, 2011: p16) to ensure the person’s wellbeing and that this added significantly to caregiver stress (Mahoney, *et al.*, 2003). For example: a qualitative study by (Feil, Lukman, Simon, Walston, & Vickrey, 2011) revealed that carers were aware of the severe outcomes that can result from diabetes including retinopathy and loss of sight but that they often found it difficult when the person they care for refuses to follow treatment instructions.

2.4.6 Interim Summary

The last section explored age related eye conditions alongside a diagnosis of dementia allowing the reader to become aware of the challenges faced by individuals and their caregivers when they face life with this ‘*double whammy*’ situation – vision loss due to eye disease plus cognitive decline. Exploration will now turn to vision loss caused by the condition of dementia itself.

2.5 Visual impairments caused by dementia

Over the age of 40 years, our eyes can develop visual problems due to the ageing process (NHS, 2018). Older people can also experience changes in sensory abilities because of neurological impairments. The link between sensory loss and cognitive function has been explored in various cross-sectional studies (Baltes & Lindenberger, 1997; Wahl & Heyl, 2003; Raina, Wong, & Massfeller, 2004; Rait, *et al.*, 2005; Roberts & Allen, 2016)

reported a direct link between impaired perception and cognitive decline suggesting that, over time, sensory deprivation leads to cognitive decline. This is an important point because people with the amnesic form of mild cognitive impairment have an increased risk of developing dementia (Ray & Davidson, 2014). A sensory risk factor example was highlighted by (Livingston, *et al.*, 2017) in a Lancet Commission report which identified a possible relationship between hearing loss and onset of dementia citing a number of cohort studies that suggested even mild levels of hearing loss may increase the long-term risk of cognitive decline. Enhancing a person's living environment, both social and physical, using person centre care and dementia design techniques may enrich their overall wellbeing through improving their ability to make sense of their world.

Visuo-perceptual difficulties have been reported in different types of dementia. For example: vascular dementia (particularly if related to stroke), and dementia with Lewy bodies. These comprise of a variety of '*visual mistakes*' including misperceptions, misidentifications, illusions and hallucinations (Alzheimer's Society (a), 2016). Types of hallucinations that can occur in people with dementia include visual, auditory, gustatory, olfactory and tactile. The most common are visual hallucinations. These phenomena are different from visuo-perceptual difficulties because they relate to things that are not present rather than mistaking or misperceiving real objects. Hallucinations in people with dementia have been reported in numerous studies, particularly AD (Murgatroyd & Prettyman, 2001). The first description of a patient with AD was reported by Alois Alzheimer who mentioned in his work not only confusion and memory loss, but also hallucinations and delusions (Grand & Feldman, 2007). Symptoms of dementia with Lewy bodies (DLB) are also characterised by prominent visual hallucinations (Holroyd, 2004).

(Jones, van der Eerden-Rebel, & Harding, 2006) have described AD as a "*simultaneous visuo-perceptual and cognitive illness*" (p3). Research has shown that people with Alzheimer's disease have difficulties with contrast sensitivity, visual attention, object and facial recognition, colour and depth perception, glare, motion perception and visual misinterpretation. For

example (Cronin-Golomb & Hof, 2004) demonstrated that people with AD showed a decline in one or more visual capacities, which were not the result of the normal ageing process.

Because people with AD may not be able to make sense of what they see or be able to explain what they have seen, (Jones, van der Eerden-Rebel, & Harding, 2006) advised that this could impact on personal behaviours, highlighting that there was a need for carers to understand that:

“...persons with AD (maybe) ‘behaving’ relatively normally in a ‘perceptually abnormal’ world (versus the more common notion of a person with dementia ‘behaving as abnormally in a normally perceived world’)” (p60).

Perrin (1997), observing 109 severely cognitively impaired persons in nine dementia units hypothesised that people experiencing advanced dementia live within a *“bubble environment”* with their visual and hearing capacities seriously compromised. By this stage of the disease, global brain atrophy has occurred. Verbal communication is minimal or impossible, so individuals are unable to explain what they are seeing or hearing. In these circumstances, care providers find it is difficult to understand how these people are perceiving their immediate surroundings and what influences could negatively or positively affect their well-being.

2.5.1 Interim Summary

The last section examined how vision problems can occur through the condition of dementia itself and how, because of lack of insight, individuals may not understand or be able to explain what is happening to them visually. **Table 4** outlines key points from **Section 2.5**.

I will now lead the reader into the main area of study namely Posterior Cortical Atrophy. The literature regarding the visual elements of this rare subset dementia will now be examined in greater detail to improve understanding around what causes the condition; recognise its specific visual manifestations and understand any current research gaps that would add to the knowledge base for this atypical dementia.

Key points from Section 2.5

Table 4: Key Points from Section 2.5

✓	People can experience changes in sensory abilities because of neurological impairments
✓	AD has been described as a simultaneous visuo-perceptual and cognitive illness
✓	People with AD have shown a decline in one or more visual capacities, which were not the result of the normal ageing process.
✓	Visuo-perceptual difficulties have been reported for other dementias including: vascular dementia (particularly if related to stroke), and dementia with Lewy bodies.
✓	People with dementia can experience a variety of ‘visual mistakes’ including misperceptions, misidentifications, illusions and hallucinations.
✓	People with dementia may not be able to make sense of what they see or be able to explain what they have seen. This could impact on personal behaviours.
✓	People with severe dementia experience severely reduced visual and hearing capabilities.

2.6 Posterior Cortical Atrophy (PCA)

2.6.1 Introduction

Posterior Cortical Atrophy (PCA), is a rare atypical dementia that presents in people in their 50s and early 60s. It is characterised by a striking decline in higher order visual-processing abilities and other cognitive impairments due to progressive atrophy in the parietal, occipital and occipitotemporal regions of the brain. Memory and language skills, at least in the early stages, remain relatively spared. This often causes a delay in diagnosis due to PCA’s unusual presentation (Crutch, *et al.*, 2012).

Although PCA has been recognised for more than three decades, until recently there was a paucity of research into the condition. An increased interest in this rare neurodegenerative syndrome has emerged in recent years with an international multi-disciplinary group of researchers forming

a working party in 2012 to raise awareness of the disease and agree a set of consensus criteria for diagnosis. (Crutch, *et al.*, 2017).

This section contains an overview of research into this condition including the history and definitions of PCA; epidemiology; neuropsychological and clinical features; and diagnostic criteria. Management of PCA (both pharmaceutical and non-pharmaceutical) currently available to improve daily life for individuals and caregivers living with PCA is also discussed. Finally, gaps in the current PCA literature are highlighted and future research areas are suggested.

2.6.2 History and definition of PCA

(Goldstein, Ivanov, & Silverman, 2011) reported that Arnold Pick, a professor of psychiatry from Prague, (better known for his discovery of one of the causes of frontotemporal lobar degeneration now named 'Pick's Disease) described a patient with symptoms indicative of PCA as early as 1902.

This was followed in 1909, by a Hungarian physician, Rezső Bálint, who reported a rare neurological condition (now named after him) which caused a patient to experience disabling visual problems due to bilateral damage of the posterior parietal lobes. A number of studies have noted the relationship between Balint syndrome and PCA citing symptoms such as simultanagnosia, optic ataxia and ocular apraxia (Husain & Stein, 1988; Crutch, *et al.*, 2012; Chechlacz & Humphreys, 2014). A description of these manifestations can be found in [Section 2.6.4](#).

The term posterior cortical atrophy, (PCA), also known as Benson's syndrome, was formally described by Benson and his colleagues in 1988. They had studied five patients aged between 54 years and 75 years who presented with slowly progressive dementia in addition to disabling higher visual disorders. Over time they all developed alexia, agraphia, visual agnosia with components of Balint's syndrome (simultanagnosia, optic ataxia and ocular apraxia); Gerstmann's syndrome (agraphia, acalculia, finger agnosia, and right-left disorientation) and transcortical sensory aphasia syndrome. Memory and insight appeared relatively preserved

until later in the condition. At that time, histopathological data was not available. Nevertheless, Benson and his colleagues judged that because the clinical presentation “*appeared unique*” with symptoms sufficiently different from AD that it should be categorized separately “*until definitive pathologic information becomes available*” (Benson, Davis, & Snyder, 1988: p793) .

In recent years, there has been a growing body of research that indicates that PCA can be attributed to a number of neurodegenerative pathologies. In most cases, the underlying primary aetiology is Alzheimer’s disease (AD) (Caine, 2004; Tang-Wai & Graff-Radford, 2011; Jellison, 2017). Indeed, PCA has been referred to as the “*visual variant of AD*” (Borruat, 2013, p406) or as an atypical variant of Alzheimer’s disease (Lee & Martin, 2004; Metzler-Baddeley, Baddeley, Lovell, Laffan, & Jones, 2010; Crutch, *et al.*, 2011). However, other degenerative causes have also been identified. For example: Lewy body disease (Crutch, *et al.*, 2017), corticobasal degeneration (Tang-wai, Josephs, Boeve, *et al.*, 2003); subcortical gliosis and, very rarely, prion disease, such as Creutzfeldt Jacob disease (Victoroff, Webster Ross, Benson, Verity, & Vinters, 1994). Findings in the last few years resulted in an emerging increased interest to regard PCA as a distinct nosological entity. Such attention culminated in the formation of an international research working party in 2012 focused on developing a consensus classification framework for the syndrome that can be used in a various clinical and research settings (Crutch, *et al.*, 2017) see [Section 2.6.7.](#)

2.6.3 Epidemiology

Because of the rare nature of the syndrome, as well as inconsistency in diagnostic criteria, the prevalence of PCA is difficult to determine. A number of researchers have suggested that PCA, like AD should be viewed as a disease of old age, (Zakzanis & Boulos, 2001; Charles & Hillis, 2005). However, the majority of studies have shown that patients with PCA present at a much earlier age than tAD usually from 50 years to early 60s (Mendez, Ghajrania, & Perryman, 2002; McMonagle, Deering, Berliner, &

Kertesz, 2006; Crutch, *et al.*, 2012; Crutch, *et al.*, 2013) although individuals in their 40s (Kas, *et al.*, 2011) and late 80s (Tang-Wai, *et al.*, 2004) have also been reported.

It has been estimated that 5% of patients seen in specialty dementia diagnostic centres have PCA (Snowdon, *et al.*, 2007) and up to 13% of individuals presenting with early onset AD have the syndrome (Koedam, *et al.*, 2010). Some studies have found women are overrepresented (Snowdon, *et al.*, 2007) while others show no gender difference (Mendez, Ghajarania, & Perryman, 2002; McMonagle, Deering, Berliner, & Kertesz, 2006). For instance, a study by Snowdon *et al* (2007) examining the cognitive characteristics of 523 AD patients showed a female bias of those presenting with PCA whereas a review by Mendez *et al* (2002) found a gender split of roughly 50-50 in a review of 69 patients with PCA. There is a paucity of prospective studies exploring disease duration although (Schott & Crutch, 2019) have suggested that PCA can have a slow progression with individuals living with the condition for more than a decade.

2.6.4 Neuropsychological and clinical features

2.6.4.1 Introduction

The early symptoms of PCA can be nebulous. There are often delays in diagnosis because of individuals being younger and the presentation of their visual or motor problems being uncharacteristic of typical dementia presentations. In many cases, a visit to an optician and/or ophthalmology consultant is first undertaken because the person is experiencing visual difficulties. Because language and insight is not usually affected in the early stages, individuals are aware of their problems and can describe the challenges they are experiencing (Nagaratnam, Nagaratnam, Jolley, & Ting, 2001; Lee & Martin, 2004; McMonagle, Deering, Berliner, & Kertesz, 2006; Beh, *et al.*, 2015; Crutch, Yong, & Shakespeare, 2016). Unlike tAD, where memory loss is frequently the first symptom observed by family members or friends during the early stages of the condition, individuals with PCA often seek help themselves because of problems reading, driving or navigating their environment (Talan, 2011). Ophthalmological examination

findings are usually normal because the symptoms are caused by visuo-perceptual and visuospatial impairments due to brain cell deterioration specifically in the posterior part of the cerebral cortex at the back of the brain where the occipital lobes are situated rather than eye diseases (Ellison, 2016).

2.6.4.2 Anxiety and Depression

Anxiety is a common neuropsychological feature in PCA. Two studies comparing the neuropsychiatric profiles of PCA and AD patients found anxiety was particularly prominent with early stage PCA alongside depression, apathy and irritability (Isella, *et al.*, 2014; Suarez-Gonzalez, Crutch, Franco-Macias, & Gil-Neciga, 2016). As an early on-set dementia, working age people with PCA may be accused of being lazy or “*malingering*” (Crutch, *et al.*, 2012, page 172). Not being believed or understood regarding the difficulties they are experiencing could, understandably, further increase stress levels. With progression of disease, activities of daily living skills and self-care become more challenging resulting in an inability to work and loss of livelihood (Benson, Davis, & Snyder, 1988). For example: a study by Shakespeare *et al.* (2015) considered the impact of visual/spatial difficulties on daily living in 42 people living with PCA and 85 individuals with tAD. A questionnaire completed by their caregivers revealed that patients with PCA had difficulty with personal self-care tasks as well as activities such as using the telephone, writing, handling money, using electrical appliances. They also reported sleep problems. In contrast, caregivers of those with tAD reported that they had difficulty in tasks requiring memory skills.

Because PCA can occur in working age individuals, as visual and perception difficulties increase, individuals eventually have to stop working. A report reviewing the literature on early onset dementia discusses loss of employment stating that this can lead to not only financial worries but also feelings of social isolation and loss of friendships for both the person with dementia and their caregivers (Collins Hunt, 2011).

“Middle-aged people with dementia face numerous psychological stressors, including uncertainty of diagnosis, coping with chronic illness, and grief related to what will be, ultimately, a fatal disease.” (Collins Hunt, 2011 page 31).

Mendez *et al* (2002) suggested that PCA patients have more insight regarding their condition than people with AD and were therefore more likely to be clinically depressed and in need of antidepressant medication.

Social isolation and depression has been linked with loneliness. (Victor, *et al.*, 2020) recently undertook a study looking at prevalence and determinants of loneliness in people living with dementia. Of the 1547 people with mild to moderate dementia questioned, 31.1 per cent reported being moderately lonely and 5.2 per cent felt very lonely. The researchers highlighted the need for more research into the field of loneliness and dementia. No research could be found within the literature which examines the impact that PCA has on loneliness, social isolation and depression although anecdotal findings from PCA support groups suggest that improving personal knowledge about the disease and gaining tips from people with the condition on how to ameliorate visual and perceptive problems can improve wellbeing (Rare Dementias Support (UCL), 2021).

2.6.4.3 Higher Cortical Visual Dysfunction

Unlike age related eye conditions that result in loss of central vision, tunnel vision, patchy or blurred vision, completely different visual manifestations can occur in people with PCA (Beh, *et al.*, 2015). Individuals might have some or all the following symptoms, which increase in severity as brain cells associated with the higher cortical visual parts of the brain deteriorate.

2.6.4.3.1 Deterioration in visual capacities

(Crutch, *et al.*, 2012) advised that individuals with PCA can experience a heightened **sensitivity to glare** from shiny surfaces. It has also been reported that people can find it difficult or unable to perceive patterns due

to impaired **contrast sensitivity** and **depth perception**. Such problems make it difficult to walk on patterned surfaces (Rogelet, Delafosse, & Destee, 1996; Cronin-Golomb & Hof, 2004). **Visual neglect** is also frequently found in people with PCA. (Andrade, *et al.*, 2010) assessed 24 people with a diagnosis of PCA and found 16 had signs of visual neglect, with the left-sided neglect being more frequent than right-sided. Such problems have important implications in everyday life as they increase the risk of falls; impact on spatial disorientation; and put people at risk of serious accidents to themselves and others if they continue to drive.

2.6.4.3.2 Achromatopsia

The person with achromatopsia becomes totally colour blind and not able to perceive colour or achieve acceptable visual acuity even in high light levels. A study comparing people with PCA and tAD by (McMonagle, Deering, Berliner, & Kertesz, 2006) found one person out of 19 subjects with PCA with achromatopsia whilst (Whitwell, *et al.*, 2007) noted that five out of 38 PCA subjects were identified as experiencing this phenomenon.

2.6.4.3.3 180-degree upside-down room tilt illusion

A room-tilt illusion consists of a clear and bright visual view of an upside-down tilt of a person's surroundings. Usually the walls and objects of the room are perceived with no distortion. The illusion is usually transitory lasting only a few minutes. A study by (Crutch, *et al.*, 2011) reported an example of a 57-year-old retired health professional who experienced this phenomenon when entering her kitchen

"It was very odd ... the whole of the room was upside down ... the bottom was on top and the top was on the bottom ... It was most peculiar ... then it was just OK...it flipped back." (p171/172).

2.6.4.3.4 Static and moving images

Akinetopsia, also known as *motion blindness*, occurs when a person can see stationary objects but cannot perceive motion. Instead they see a series of still images (Crutch, *et al.*, 2012). (Tsai & Mendez, 2009) described this occurrence in two individuals with PCA: a 71-year-old woman who was

having double vision with *“leftward moving objects perceived as two or more successive images occurring side by side”* and a 61-year-old man who experienced akinetopsia when fingers or objects were moved to the left. He also described *“a sensation of double or multiple images when objects were moved in a leftward direction”* (p731). Conversely, some people with PCA are unable to detect stationary objects but can grasp them when they are moving. For example: Midorikawa, Nakamura, Nagao, & Kawamura, (2008) described a patient with middle-stage PCA, whose prominent symptom was the inability to perceive static objects. He could not detect a motionless tennis ball, even when it was in front of him. In contrast, he could find and grasp the same ball with ease when it moved.

2.6.4.3.5 Reading difficulties

Reading difficulties are common symptoms in PCA, occurring in 80 per cent of all cases (Mendez, Shapira, & Clark, 2007). **Alexia** describes someone who has lost the ability to read or understand words, sentences and in some case letters. It is also called *word blindness* or *acquired dyslexia*. It can occur in people who have suffered damage to the left side of the brain (in the occipital and temporal lobes). A review by (Crutch, *et al.*, 2012) noted that several processes limited reading skills in PCA: **reverse-size phenomena** where people can perceive small but not large print; **visual disorientation** where a person is unable to track a line of text or is losing their place on the page when they are trying to read (Benson, Davis, & Snyder, 1988); and **visual crowding** where letters can be recognised in isolation but not when they are mixed within larger chunks of prose (Crutch & Warrington, 2010). Difficulties can also happen through **ocular motor apraxia** where the person finds it very difficult or even impossible to control their eye movements. A case study by Tondo, De Marchi, Terazzi, Sacchetti, & Cantello (2017) describes a 62-year-old female with PCA who presented with all the above difficulties, succumbing over the life of the study to a rapidly deteriorating dementia.

2.6.4.3.6 Agnosia

Agnosia is the inability to process sensory information (Biran & Coslett, 2003). Lissauer introduced the concept of **apperceptive agnosia** in 1890 (Warrington & James, 1988). This causes a person great difficulty in their ability to recognise visually presented information (Goethals & Santens, 2001; Giovagnoli, *et al.*, 2009) although they can recognise objects from touch or hearing. (Gardini, *et al.*, 2011) described a gentleman who could recognise simple objects but had difficulty in identifying multipart objects. Similarly, (Mizuno, Saror, Liccione, Battelli, & Campo, 1996) described a 53-year-old man who when performing visual tasks, showed severe impairment of visual perception with the naming of objects in pictures. However, he was able to name objects based on verbal description.

With **associative agnosia**, an individual can copy or match objects but is unable to identify simple objects even with apparent perception and knowledge of them. (McCarthy & Warrington, 1986) described a 77-year-old, previously well, retired shopkeeper who had woken one day to discover he could no longer read the paper. Gradually his visual difficulties became worse and when tested was found to have lost his ability to recognise the meaning of visually presented objects and found it impossible to name or point reliably to a named object although he was reasonably able to recognise and name visual representations of actions.

If a person is experiencing **environmental agnosia** they struggle to recognise surrounding even when they are familiar to them, such as their own home or room. They also have difficulty learning routes so get lost easily. (Landis, Cummings, Benson, & Palmer, 1986) described a study of sixteen PCA patients who, despite having intact verbal memory, cognition and perception, had lost the ability to recognise familiar settings.

Josef Gerstmann was the first to define **finger agnosia** in 1924. The syndrome causes an individual to be unable to distinguish, name or recognise their own fingers as well as the fingers of other people and other depictions of fingers. A study by (Davis, Trotter, Hertz, Bell, & Dean, 2012) of 28 people with AD compared with 28 healthy subjects found that those

with AD demonstrated significantly higher incidence of finger agnosia in both hands. It was suggested that this was due to impairment of the parietal area of the brain.

Mirror agnosia has been reported in people with dementia and PCA where the person does not recognise themselves in a mirror (Breen, Caine, & Coltheart, 2001). This can cause great upset to an individual. The syndrome was reported by (Ramachandran, Altschuler, & Hillyer, 1997) who described four stroke patients with right parietal lesions who, during testing, complained that the object was *“in the mirror”*, *“outside my reach”* or *“behind the mirror”* (p645). More recently (Chandra & Issac, 2014) described five elderly patients with moderate stage posterior dementia who presented with mirror agnosia. *Case One* on viewing herself in the mirror became quite hostile to the image, quarrelling with the woman in the mirror and asking her who she was. *Case Two* recognised her clothes and ornaments but accused the mirror image of stealing her clothes and jewellery. *Case Three* presumed the image in the mirror was a thief and kicked it. *Case Four* declared the image an old friend and *Case Five* believed the image was an old neighbour he knew quite well.

Simultanagnosia results in a person only being able to perceive individual elements of a visual scene but being unable to integrate them all together to form a whole scene (Dalrymple, Barton, & Kingstone, 2013). Neitzel, et al. (2016) investigated 12 patients with PCA, all having homogenous lesions in the parieto-occipital cortices as well varying grey and white matter loss. All individuals presented with severe deficits in simultaneous perception. Although individual short-term memory was preserved, there was a specific slowing of visual processing speed in all cases. The study found that more severe slowing of processing speed resulted in more serious simultaneous perception tasks. An example of simultanagnosia in a male music teacher is described by Sacks (1986):

“I opened out a copy of the National Geographic Magazine, and asked him to describe some pictures in it. His response was very curious. His eyes would dart from one thing to another, picking out tiny features ... a striking

brightness, a colour, a shape – but in no case did he get the scene-as-a-whole.” Page 11.

(Boucart, Calais, Lenoble, Moroni, & Pasquier, 2014) assessed 14 patients with tAD and 6 patients with PCA alongside 25 healthy participants. People with PCA found it more difficult to detect the target object within a natural scene than healthy participants or people with tAD. (Shakespeare, *et al.*, 2013) also examined scene perception in 13 patients with PCA who showed similar perceptual deficits.

In **prosopagnosia**, also known as ‘**face blindness**’, the person is unable to recognise familiar people from their face. This includes family members and friends as well as famous people. It is thought to be the result of damage to the right fusiform gyrus, the part of the brain that is responsible for facial perception and memory (NINDS, 2017). Again, Sacks (1986) gives an excellent example of this visual disfunction describing once again the visual difficulties experienced by the aforesaid music teacher:

“On the walls of the apartment there were photographs of his family, his colleagues, his pupils, himself ... by and large, he recognised nobody: neither his family, nor his colleagues, nor his pupils, nor himself ... He recognised a portrait of Einstein, because he picked up the characteristic hair and moustache ... In the absence of ‘obvious markers’ he was utterly lost.” (p14)

2.6.4.3.7 Complex visual hallucinations

Complex visual hallucinations are vivid phenomena often involving animals and human beings in bright colours and dramatic settings. (Manford & Andermann, 1998) advised that the emotional reaction could evoke feelings of pleasure through to extreme fear. It has been reported that visual hallucinations occur in 25 per cent of people who met the criteria of posterior cortical atrophy (Josephs, *et al.*, 2006). In a voxel-based morphometry study of 59 individuals with PCA, 13 patients were experiencing vivid hallucinations, the majority starting four years following PCA diagnosis. These hallucinations were vivid, recurrent, spontaneous and had been present for many years. Progressive damage to

thalamocortical connections was suggested with a consideration that these individuals should also be given a diagnosis of DLB.

2.6.4.3.8 Palinopsia

Individuals with PCA have described seeing images after their corresponding stimulus has been removed. This visual disturbance is called **palinopsia** or *after-images*. The source of such occurrences was conventionally thought to be caused by the retina, but it is now known they occur through a cortical process. Gersztenkorn & Lee (2014) undertook a literature review of all cases of palinopsia. They subdivided the symptoms into two clinically relevant groups. **Illusory palinopsia** which is usually caused by migraine, head trauma or drugs which describe afterimages which are indistinct and blurry; and **hallucinatory palinopsia** which are high clarity and longer lasting. These are caused by epileptic seizures or posterior cortical lesions. (Chan, Crutch, & Warrington, 2001) describe a 64-year-old woman diagnosed with AD and probable PCA who explained that after making her bed with red sheets, she noticed her hands were coloured green. The colouration lasted up to a minute before her hands gradually resumed their normal colour. Following an MRI scan, they discovered that the patient had atrophy of the V1 layer of the visual cortex and this dysfunction was generating the colour after-images.

2.6.4.3.9 Optic ataxia

A component of Balint's syndrome, **optic ataxia** causes a person to be unable to reach or grasp objects using visual information. This is caused by damage to the posterior parietal cortex and so can affect people with PCA. It affects the visual field opposite the damaged hemisphere (Khan, Pisella, Delporte, Rode, & Rossetti, 2013). This condition was reported by Holmes (1916) when describing a World War One patient "*when as to touch or grasp my hand or a pencil held in front of his eyes, he groped wildly for it and ... brought his hand beyond it when it was within his reach*" (described by Andersen, Anderson, Hwang, & Huaschild, (2014, p3) in a review of the condition).

2.6.4.3.10 Dressing and constructional Apraxia

Dressing and constructional apraxia occurs because of visuospatial deficits. This causes an individual to find it very difficult or impossible to perform tasks or movements. Individuals find it problematic to dress often putting clothes on back to front or upside down. Drawing a clock face or copying diagrams becomes more and more difficult with PCA deterioration (Benson, Davis, & Snyder, 1988).

2.6.5. Other symptoms of PCA

In addition to higher visual dysfunction as described above, other non-visual deficits in PCA are common. These include:

2.6.5.1.1 Acalculia

Acalculia describes the loss of ability to perform simple mathematical calculations. Loss of mathematical abilities has been identified as a symptom of tAD (Martin, *et al.*, 2003) but it has also been reported widely as a symptom of PCA (Benson, Davis, & Snyder, 1988; Schott & Crutch, 2019; Gonzalez, Hoffman, & Crutch, 2019). For example, all five cases reported by Benson *et al* (1988) had lost the ability to perform simple arithmetic even when given paper and pencil to assist them. One individual, a 59 year old bookkeeper, recounted that his problems arose first by having difficulty following a line of print followed a year later having *“problems in calculations, even in keeping the family check book”* (page 790) resulting in him losing his job. A recent study by Harding *et al*, (2018) identified problems in handling money or poor financial skills as negative factors when undertaking daily living skills.

2.6.5.1.2 Anomia and Agraphia

Anomia is a form of **aphasia** and describes the ability to use an everyday object but not be able to recall its name whilst **Agraphia** is the inability to write words, including a person’s own signature. (Sacks, 2010) recalled a lady called Lilian, a gifted musician, diagnosed with PCA who over time developed anomia, and how she dealt with her difficulties:

“When I showed her some kitchen matches, she recognised them at once, visually, but could not say the word ‘match’ saying, instead “That is to make fire” She was well aware of these difficulties, and of her strategies dealing with them “When I can’t say something”, she explained, I circumscribe.” Page 26

Schott & Crutch (2019) described a general practice nurse who began to have difficulties writing patients notes or transcribing information to a computer. Likewise, Goethals & Santens (2001) reported two cases of PCA, the first a 59 year old teacher lost the ability to draw and write resulting in loss of employment; and second, a 55 year old housewife who lost the ability to write and found copying simple figures impossible.

2.6.6 Differences between tAD and PCA

The Global Deterioration Scale describes the progress of dementia, (specifically tAD) using stages (Reisberg, Ferris, de Leon, & Crook, 1982). Crutch, Walton, Carton, & Shakespeare (2017) applied this stage approach to show the progression of PCA. They emphasised that this progression, as with tAD, is different for every individual depending where damage in the brain occurs, subsequently influencing the visual problems experienced. It is interesting to compare the differences of each stage of PCA against each stage of tAD because it highlights the variation of symptoms of the two dementia disorders. These are described in [Table 5](#) where the different presentations of the two conditions are set side by side. [Appendix Two](#) shows a comprehensive list of the symptoms that occur at stages of PCA (Crutch, Walton, Carton, & Shakespeare, 2017).

Table 5: Differences in presenting symptoms of tAD and PCA

Stages	Symptoms in tAD	Symptoms in PCA
Stage 1 (No cognitive decline)	The person does not experience any memory or other cognitive problems	The person does not experience any visual problems, memory or other cognitive problems.
Stage 2 (very mild cognitive decline)	<p>The person experiences subtle difficulties such as subjective symptoms of memory deficit, such as:</p> <ul style="list-style-type: none"> • Forgetting the location of familiar objects • Forgetting previously well-known names. 	<p>The person experiences subtle difficulties with some complex visual tasks such as:</p> <ul style="list-style-type: none"> • a loss of confidence in their driving ability • problems using a computer • difficulties with reading, writing, spelling and/or arithmetic • difficulty recalling and writing down telephone numbers • difficulty assembling novel devices/objects • The person may experience vertigo or other balance problems • may experience difficulties finding the right words when speaking or may experience odd visual sensations (e.g. occasional changes or washes of colour in the centre or periphery of their vision). <p>Eye tests reveal no indication of ophthalmological problems.</p>
Stage 3 (Mild Cognitive decline)	<p>Difficulties acknowledged by person, family and co-workers, such as:</p> <ul style="list-style-type: none"> • Early, clear-cut memory • getting lost, • poor work performance, • problems with word or name finding, • misplacing objects of value, • poor retention of written material 	<p>Visual difficulties are acknowledged both by the person and their friends, family and co-workers. Healthcare professionals familiar with young onset and atypical dementias may recognize these problems:</p> <ul style="list-style-type: none"> • Difficulty seeing things or only seeing part of something • General 'clumsiness' • Difficulty finding things in a handbag, cupboard, etc. – • Difficulties dressing and performing other activities of daily living that require spatial judgement • Inability to tell the time from a clock or watch • Reading and writing difficulties • Mild memory problems for recent events • Mild difficulty retrieving the correct word in conversation

<p>Stage 4 <i>(Moderate cognitive decline – early dementia)</i></p>	<p>Person presents with clear cut memory deficits, such as:</p> <ul style="list-style-type: none"> • poor knowledge of current events • difficulty remembering details of personal history • impaired concentration doing serial subtractions • inability to handle finances or travel to family locations. 	<p>Individuals require significant help with everyday activities such as cooking, dressing and using appliances. The person retains a clear sense of purpose but is frustrated by frequent, small visual problems:</p> <ul style="list-style-type: none"> • Inability to read labels on food packets – • Problems discriminating coins – • Inability to tell the time from an analogue or even digital watch • Dissociation in ability to walk, sit down and stand up • Being able to see some things but not others • Cannot read but can still follow or at least get pleasure from TV or the cinema • Getting lost within the home. • An inability to identify even very familiar faces • Resorting to eating with one’s hands, or being fed • increasing word finding difficulties
<p>Stage 5 <i>(Moderately severe cognitive decline - middle dementia)</i></p>	<p>Person has serious memory deficits and can no longer function without some assistance.</p> <ul style="list-style-type: none"> • Unable to recall major aspects of current life (e.g., address, telephone number, names of family members). • May need assistance dressing, but still independent in eating and toileting. 	<p>Many individuals are now registered as blind. They may now:</p> <ul style="list-style-type: none"> • Require help with most or all everyday tasks, including toileting and personal care • Experience spatial problems not limited to vision • Require support and/or supervision when walking • Episodic memory and word finding difficulties may also now be prominent • Some people may have problems with multiple senses • Increased sensitivity to pain • Become more sensitive to the cold • Sense of hearing may change • Feelings of imbalance or instability

<p>Stage 6 <i>(Severe cognitive decline – middle dementia)</i></p>	<p>Person has extremely serious memory deficits including:</p> <ul style="list-style-type: none"> • Occasionally forgetting name of primary caregiver (e.g., spouse). • Is largely unaware of recent personal events. • Needs substantial assistance required for activities of daily living and travel to familiar locations. • Often unaware of surroundings (e.g. year or season). • Can still recall his or her own name and distinguish a familiar face. 	<p>Cognitive changes are more global in nature, with multiple aspects of perception, memory, language, attention and decision-making abilities affected. Vision remains the most pronounced impairment, but in most individuals, there are widespread impairments that would be recognised as dementia. They may:</p> <ul style="list-style-type: none"> • Experience changes in sleep patterns • Have increasingly frequent trouble controlling their bladder or bowels – • Become quiet and withdrawn • Experience behaviour and personality changes • Have difficulty communicating • Experience continued deterioration of sensory functions
<p>Stage 7 <i>(Very severe cognitive decline – Late dementia)</i></p>	<p>Person has catastrophic memory deficits including:</p> <ul style="list-style-type: none"> • Being unable to recognise familiar faces. • Verbal ability limited to less than 5 words. • Incontinent of urine and stool. • Total functional dependence • unable to walk. 	<p>In the late stages, the problems experienced by people with PCA may resemble typical AD more closely than at any stage including:</p> <ul style="list-style-type: none"> • Loss of ability to respond to their environment • Unable to carry on a conversation • Unable to control movement. • Total functional dependence • Unable to walk
<p>End of Life</p>	<p>Although Alzheimer’s disease and other degenerative diseases are life shortening illnesses, another condition or illness (such as pneumonia) may cause the person’s death. Depending on the circumstances, ‘Alzheimer’s disease’ or similar may be entered on the death certificate as the sole or main cause of death, or as a contributing factor.</p>	<p>Although Alzheimer’s disease and other degenerative diseases [such as PCA] are life shortening illnesses, another condition or illness (such as pneumonia) may cause the person’s death. Depending on the circumstances, ‘Alzheimer’s disease’ or similar may be entered on the death certificate as the sole or main cause of death, or as a contributing factor</p>
	<p>+ (Reisberg, Ferris, de Leon, & Crook, 1982)</p>	<p>++ (Crutch, Walton, Carton, & Shakespeare, 2017)</p>

2.6.7 Diagnostic criteria

The classification used to clinically diagnose PCA has been varied. Some researchers viewed PCA as a 'visual atypical form of AD', whilst others determined that multiple pathologies could trigger the condition. For instance, Mendez (2004) and Tang-Wai, *et al.*, (2004) concentrated on visual impairment, whereas Renner, *et al.* (2004), focused on a broader definition which included language, movement and somatosensory dysfunctions. Terminology also differed. PCA sometimes being labelled a descriptive clinical syndrome and by others termed PCA as a diagnostic label.

In 2012, an international multidisciplinary working group was set up to develop consensus recommendations for diagnosis and classification of the syndrome which would address these inconsistencies. Their goal was to:

“review, revise, and complement existing single-center diagnostic criteria to represent multidisciplinary and multicenter experience and knowledge”

(Crutch, *et al.*, 2017) page 872

The group published a report detailing a three level classification framework of PCA in 2017 defining its core clinical, cognitive and neuroimaging features and exclusion criteria. (See Table 6). The group suggested that such a framework helped clinicians could assist in speedier diagnosis and enable people to obtain the specific help they need to manage the condition (Crutch, *et al.*, 2017).

Table 6: PCA Classification Framework

PCA Classification Framework	
Classification Level One:	Details the core feature of the PCA clinic-radiological syndrome.
Classification Level Two:	At this level, individuals are split into two groups. Those who solely meet the criteria for PCA (PCA-Pure), and those that exhibit additional neurodegenerative syndromes such as DLB or CBS.
Classification Level Three:	This level provides disease level descriptions of PCA reflecting evidence of underlying pathology. For example: PCA attributable to AD (PCA-AD); Lewy Body disease (PCA-LBD); Corticobasal degeneration (PCA-CBD) and prion disease (PCA-prion)
<i>Information taken from (Crutch, et al., 2017)</i>	

2.6.8 Management of PCA

NHS health policies have stressed that **living well with dementia** is a key outcome in the care of people with dementia (Department of Health (a), 2015) (Department of Health (b), 2015). The idea of wellbeing through positive mental health was espoused as far back as the 1950s by (Jahoda, 1958). (Power, 2014) suggested that in the case of a person living with dementia, using a person-centred value base, where comfort, attachment, inclusion, identity and occupation, espoused by Brooker (2007), is met, will help maintain positive wellbeing, whilst disregarding those elements would result in ill-being.

In 2007, NICE/SCIE, (National Collaborating Centre for Mental Health , 2007) published guidelines on supporting people with dementia and their carers advocating a holistic approach to care. The main causes of dementia were described but at that time, little was known about PCA and the specific challenges resulting from this visual-variant of AD were not

considered. Guidance published 2016 also did not include PCA in its updated documents. (NICE, 2016).

The descriptions of the visual-perceptual and visual-spatial disorders of PCA previously detailed can cause serious disability. (Metzler-Baddeley, Baddeley, Lovell, Laffan, & Jones, 2010) described that such impairments:

“can leave the patient behaving like a blind person” p36.

With the progression of PCA symptoms, most people find it impossible to continue in employment and become dependent on caregivers to maintain their quality of life and as can be viewed in **Table 5** when the person is experiencing the end stages of the disease, the differences between tAD and PCA are minimal (Crutch, Walton, Carton, & Shakespeare, 2017). The importance of successful management of the condition was emphasised by (Pratchett, 2015) who lived with PCA for 10 years. He advised that he was able to lead a relatively normal life for quite a long time through the support of his family and friends making allowances for his disabilities.

Web page information about PCA has been made available to the general public by a national dementia charity and UCL which offer support to people with the condition and their caregivers (Alzheimer's Research UK/Rare Dementia Support, 2017) as well as the Alzheimer's Society (Alzheimer's Society, 2017).

2.6.8.1 *Pharmaceutical Management of PCA*

Unfortunately, no specific medical treatments are currently available to treat PCA. Schott & Crutch (2019) have suggested that pharmacologic treatments should focus on the pathologic substrate of PCA. For instance, if PCA is due to AD, patients may possibly find medications such as acetylcholinesterase inhibitors or memantine helpful (UCAF Memory and Aging Center, 2014). Individuals with PCA who experience anxiety and depression might be prescribed antidepressant medication (Schott & Crutch, 2019).

2.6.8.2 *Non-pharmaceutical Management of PCA*

With no specific pharmaceutical options for PCA, more holistic approaches to managing the condition are being developed which consider both the physical and emotional needs of people with PCA and their caregivers.

2.6.8.2.1 *Improving the physical environment*

Physical and psychological distress can be caused by negative environmental settings (Power, 2014; Rahman, 2014). People with PCA live in different care settings, but in the early to moderate stages of the condition, the majority of individuals are likely to live in their own homes. It is therefore important to make the home *'PCA friendly'* by optimising the physical environment to take account of any visual and/or perceptual problems experienced in order to maximise independence, autonomy and wellbeing for as long as possible.

The increase of specialists designing for dementia over recent years has been welcomed (Lawton, 2001). In a review of the empirical research on design and dementia, (Day, Carreon, & Stump, 2000) concluded that

“For the greatest impact, design professionals and researchers must continue to educate administrators and families on the potential role of environmental design for improving quality of life in a comprehensive way. These recommendations, if implemented, will ensure continued progress in the study and design of therapeutic environments for people with dementia.” Page 413

The majority of reviews of dementia design have focused on specialist facilities rather than individual home environments (Day, Carreon, & Stump, 2000; Fleming, Crookes, & Sum, 2007). Although good practice guidelines for people with dementia living in domestic care settings have also been published (Dementia Services Development Centre, 2010).

Whatever the setting, **Table 7** sets out therapeutic design principles that can enhance a person's quality of life. Such principles should:

1	Maintain and sustain personal identity
2	Compensate for disability
3	Maximise independence
4	Enhance self-esteem and confidence
5	Aid orientation and understanding

(adapted from (Marshall, 1998) and (Pool, 2010))

Good practice guidelines for people with typical dementias and vision loss have been reported (Bowes, McCabe, Dawson, & Greasley-Adams, 2014). Some suggested areas may also be helpful for ameliorating some of the symptoms of PCA. These include:

2.6.8.2.1.1 Light and lighting

An important feature of the visual environment in supporting people with dementia is the source and quality of light (Calkin, 1997; McNair, Cunningham, Pollock, & McGuire, 2013). Domestic housing often has lighting levels that are often too low for older people but by increasing the number and wattage of lights, a clearer view of the environment will be achieved. Hallways, stairs and entrance foyers need to have balanced lighting to enable a person's eyes to adapt more easily as they move from one area to another (Campbell, 2005). Natural daylight should also be optimised by ensuring windows are clean, opening blinds and curtains or moving furniture that might be blocking light (Dementia Services Development Centre, 2010).

2.6.8.2.1.2 Colour and Contrast

Designing a colour scheme which minimises the use of the blue-green has been recommended (Dunne, 2004) as studies have reported a deficit in

older people and people with dementia within this colour range (Brawley, 2001; Calkins, 2002). Using set colours for specific rooms or fittings may help with recognition (Pool, 2010). For example, toilet doors having the same strong distinctive colour which contrasts with the surrounding wall with the addition of clear signage using words and pictures may aid recognition and reduce incontinence. Signs should be placed just below average height, so individuals are attracted to them. Conversely, painting a door leading to a hazardous area the same colour as the wall can deter access.

The importance of contrast has been emphasised in several design publications (Utton, 2007; McNair, Cunningham, Pollock, & McGuire, 2013). For instance, contrasting colouring flooring could help distinguish the furniture from the floor. It is possible to purchase tools that scientifically identify contrast levels. However, a simpler and cheaper option would be to take a black and white photograph of a room and see how easily the furniture stands out against its surroundings (Dementia Services Development Centre, 2010).

2.6.8.2.1.4 Glare and reflections

A highly polished floor could be interpreted as water (Utton, 2007) and could prove hazardous to an individual with PCA making them prone to falling or refuse to walk across the area and becoming agitated or distressed. Reflections in mirrors, TVs or windows can also cause anxiety and agitation if the person does not recognise themselves or may think there is someone else in the room, so a cover or curtains can be useful. On the other hand, partially glazed entrance doors or cupboard doors can be helpful as they will enable the person to see what is in the cupboard or the room they are about to enter (DSDC, 2013).

2.6.8.2.1.5 Perceptual difficulties

Patterned wallpaper and carpets can be problematic. Patterned carpets may be perceived as being at different heights and individuals may try to step over them leading to a risk of falling. A dark rug or mat may be perceived as a hole and cause anxiety. Using consistent plain colour matte

surfaces can aid individuals to walk more safely (Dementia Services Development Centre, 2010).

2.6.8.2.1.6 Assisted Technologies.

The literature shows that assisted technologies improves the safety of people with dementia particularly around falls risk and accidents (Brimsa & Oliver, 2019). Assisted technologies (AT) are also used by people with visual impairments to help them with their every day activities (RNIB, 2021). Two studies were found that focused on the specific visual and perceptual needs of people with PCA with regard to helping individuals improve their reading capacities. Yong, Rajdev, Shakespeare, Leff, & Crutch (2015) examined the reading difficulties in 15 people with PCA and 6 with tAD using two software reading aids which helped ameliorate eye fixations and eye movements to improve reading ability and Suarez-Gonzalez, et al.(2019) tested a reading app on 20 individuals with PCA. They found that the tool improved subjective reading enjoyment.

2.6.8.2.1.7 Good practice guidelines

(Bartlett & McKeefry, 2009) asserted that care home inspectors should be provided with a good practice checklist to ensure that the needs of such residents with sensory disabilities are addressed and called for National Dementia Strategy leaders to fund initiatives to increase the knowledge base around the needs of people with sensory problems and dementia. Such checklists should also be available to informal caregivers supporting their loved ones in the home environment. Studies show that staff in dementia care services can lack the time and understanding to respond adequately to sensory problems (Lawrence, Murray, Ffytches, & Banerjee, 2008).

Environmental design guidelines have been published regarding the visual needs of people with dementia (Goodman & Watson, 2010; Greasley-Adams, Bowes, Dawson, & McCabe, 2016). However, these focus on people with age-related eye disease rather than individuals with visual difficulties due to dementia, such as those with PCA. Although useful to some extent, particularly regarding lighting and colour contrast issues, the

specific visual manifestations experienced by people with PCA are not being fully addressed in current dementia design guidelines.

2.6.8.3 Cognitive rehabilitation and psycho-education training

In a study comparing 32 PCA and 71 tAD individuals, (Shakespeare, Yong, Foxe, Hodges, & Crutch, 2015) highlighted the significant impairment in daily living skills in people with PCA including functional deficits such as using the telephone, using electrical appliances, reading and writing, eating and drinking, making hot drinks and problems with calculation. They also were shown to sleep more during the day.

Because of the lack of effective medication to ameliorate such challenges, non-pharmacological interventions may be appropriate, particularly in the early to moderate stages of PCA.

Cognitive training (CT) aims to improve or maintain a person's cognitive abilities by using guided practice on structured tasks over a set period of time. A recent intervention review was undertaken by Bahar-Fuchs, Martyr, Goh, Sabates, & Clare (2019) to evaluate cognitive and non-cognitive outcomes for people with mild to moderate dementia and their caregivers. The team reviewed 33 trials conducted between 1988 and 2018 and concluded that CT was probably associated with small to moderate effects on global cognition and verbal semantic fluency at end of treatment, and that these benefits appeared to be maintained in the medium term. However, they cautioned that future studies should include longer term follow up to better assess positive outcomes.

Four case studies were found in the literature which focused on cognitive rehabilitation and/or psychoeducation for people with PCA.

The first described a 64 year old male who took part in a five week intervention programme which included psychoeducation, cognitive exercises and compensatory strategies for improving activities of daily living (ADL) identified by the patient. These were to gain an understanding of PCA and its symptoms; increase his ability of finding of objects in his immediate vicinity; improve the difficulties he was experiencing pouring

drinks; and being able to understand written messages left by his wife. On completion of the rehabilitation programme both the patient and his wife felt that improvements had been made in everyday functioning. The researcher also identified subtle positive changes. Nevertheless, the researcher predicted that these improvements may only be temporary and functioning might decrease again over time due to cognitive decline (Roco, Gleichgerrcht, Torralva, & Manes, 2010).

Weill-Chounlamounry, *et al.*, (2012) used a six-month multidisciplinary programme of physiotherapy, occupational therapy and speech therapy with a 60 year-old female patient to attempt to strengthen her daily living abilities. Following the programme she was able to read (a skill she had previously lost) and felt confident in using the underground and visiting museums. They suggested that this multidisciplinary approach appeared beneficial in the absence of further cognitive decline.

A French psychoeducation study by Videaud, Torny, Cartz-Piver, Deschamps-Vergara, & Couratier (2012) reported by (Weill-Chounlamounry, Alves, & Pradat-Diehl, 2016) included four individuals with PCA and their caregivers. Over one year, the participants took part in six two-hour sessions every two months. These sessions included:

“(1) information about the disease to enhance the understanding of PCA subjects and their caregivers; (2) an assessment of repercussion of PCA on the activities of daily life (3) a speaking time; (4) use of technical aids to improve the quality of life; and (5) strengthening the knowledge and information about available help (financial and human aids).” (Weill-Chounlamounry, Alves, & Pradat-Diehl, 2016: pages 197 and 198).

No changes in quality of life for the individuals with PCA were recorded. However, a reduction of anxiety by caregivers was noted.

(Alves, *et al.*, 2015) reported modest improvements particularly in verbal memory, attention and psychomotor abilities in a 57 year-old patient with PCA who undertook 60 hours of intensive rehabilitation including reality orientation, cognitive stimulation and cognitive training exercises with the addition of psychoeducation about their condition. However, they stressed

that larger controlled studies needed to be undertaken to assess any potential efficacy of the programme.

A case study of a 69 year-old woman with PCA named DD was described by (Bier, *et al.*, 2019). DD wanted to improve her ability to play cards with her grandchildren and lay the table at mealtimes. Researchers used a cognitive rehabilitation programme using *errorless learning* and compensatory aids.

The only improvement noted was her ability in laying the table using a placemat showing where items should be placed (e.g. plate, fork, knife, glass). However, the researchers felt that their study did not contain enough sessions to be able to determine the long-term efficacy of the intervention.

To date, this field remains understudied for people with PCA (Weill-Chounlamountry, Alves, & Pradat-Diehl, 2016). The provision of more holistic approaches to care need to consider both the physical and emotional needs of people with PCA and their caregivers to improve personal wellbeing including both psychological support and practical solutions.

2.6.8.4 The subjective experience of living with PCA

(O'Connor, *et al.*, 2007) highlighted the need to fully understand the subjective experience of the person with dementia arguing the need for a multi-dimensional, interdisciplinary and comprehensive vision of dementia highlighted by (Downs, 2000). The increasing involvement of patients and users in service development has been recognised and such advances give opportunities for people living with dementia to also give their views

One implication resulting from the development of person-centred care, has been an understanding amongst both academia and health and social care providers that many people with dementia can talk about their experiences. There is a growing body of empirical research which proves this (Goldsmith, 1996; Chester & Bender, 1999; Peterson, 2004; Allan & Killick, 2008; Stokes, 2010; Whitman, 2016) as well as an increasing number of books written by people with dementia about their individual

perspectives of living with the condition (Davis, 1989; McGowan, 1993; Bryden, 2005; Kessler, 2007). In addition to academic and professional study, the voices of people with dementia are being heard via social media using blogs and YouTube as well as storylines in soaps and films (Amis, 2001; BBC, 2007; MHA, 2018). Strategies for improving the care of people with dementia for England have also highlighted that quality of life outcomes for dementia should be determined by people living with the condition as well as their formal and informal caregivers. (Department of Health, 2009) (Department of Health, 2010) (DH/SCLGCP/SCP/OP&D, 2010) (Department of Health, 2013) (Department of Health (a), 2015) (Department of Health (b), 2015) (Department of Health and Social Care, 2016).

Research to understand the subjective experiences of living with PCA is lacking. To date, only two studies were found that explored the subjective experience of PCA. The first reported the lived experiences of people living with the condition suggesting that further research exploring this atypical dementia would be worthwhile (Harding, *et al.*, 2018). The second qualitative study gathered opinions from health and social care practitioners regarding the needs of people with PCA and what training they needed in order to support such individuals (McIntyre, *et al.*, 2018).

2.6.8.5 Wellbeing of caregivers

(Alzheimer's Society, 2020) reported in the UK, approximately 770,000 were caring for someone with dementia. This numbers is probably higher as individuals sometimes do not class themselves as a caregiver but a husband/wife or child supporting a loved one. The importance of maintaining the wellbeing of caregivers has been emphasised as an important consideration (Strategy and Projects Team, Care and Transformation, 2018).

Caring for someone with dementia can be emotionally and physically exhausting. (Alzheimer's Research UK, 2019) noted that caring for someone with dementia will inevitably change the dynamics of the relationships between the person with dementia as well as the wider social

network of friends and family. Caregivers can become socially isolated which in turn impacts negatively on psychological and physical health.

As shown in **Table 8**, practical ways to build up the resilience of caregivers own health and wellbeing have been suggested:

Table 8 Practical suggestions to improve caregivers health and wellbeing:

<ul style="list-style-type: none">• Looking after yourself by talking to your GP and registering as a carer in order to get information and free health checks and flu vaccinations
<ul style="list-style-type: none">• Involving other people by accessing professional support from local authority, housing and care services and personal support from friends and family members.
<ul style="list-style-type: none">• Using respite services to take a break from caring.
<ul style="list-style-type: none">• Eating well
<ul style="list-style-type: none">• Keeping physically and mentally active
<ul style="list-style-type: none">• Making time for your own hobbies
<ul style="list-style-type: none">• Getting enough sleep
Adapted from (Parkinson, Carr, & Rushmer, 2016) and (Alzheimer's Society, 2019)

Unfortunately, a recent survey found that, where support from professional services was received, communication was often poor and people were asked to repeat their stories when meeting new professionals. The majority of survey participants had not received a carers assessment and in many regions of the UK over 80% of people had not had social services support to take a break (Alzheimer's Society, 2020).

A recent study by (Chang, Lorenz, Phillips, Peng, & Szigeti, 2020) found high levels of fatigue amongst 43 caregivers of individuals with dementia that was greatly influenced by sleep quality calling for more studies in this area in order to gain insights into ways to caregiving experiences.

Minimal studies concerning the caregivers of people with PCA have been conducted. (McIntyre, et al., 2018) interviewed health and social care practitioners regarding the needs of people with PCA and their training

requirements. Anecdotal evidence from a webpage on PCA suggested that informal caregivers can find it challenging to understand some of the behaviours of the person with PCA and that friends and family often do not understand their needs and experiences due to a lack of services available to help them learn about the condition (Hinds, 2021). To date, there is a paucity of research examining the subjective experiences of informal caregivers of individuals with PCA.

2.6.9 Gaps in the literature

At first glance, the literature shows positive progress regarding a clearer understanding of PCA and much research is now taking place regarding the bio-medical causes of the condition. However, to date, considerations for improving dementia care practices have focused on the needs of the general dementia population and the wellbeing of those that do not fall into this 'typical' dementia bracket still needs to be addressed. For example, it appears that individuals with sight loss due to the condition of dementia, such as those with PCA, have been neglected and as a result the experiences of this population is poorly understood and underrepresented.

It could be reasoned that this atypical presentation of AD is rare and so does not justify in-depth study. On the other hand, the prevalence of such a presentation should not of itself predetermine the quantity or quality of care a patient should receive. Rarity of presentation may also be a case of inability to diagnose due to unfamiliarity with the condition (as indicted in [Section 2.6.1](#)). PCA is an example of such a case, so deeper research would hasten better diagnosis, possibly leading to an increase in recorded prevalence. Arguably, therefore, the rarity suggests that understanding of the condition should be researched more deeply as those people with such a rare condition and their caregivers have fewer sources of expertise to draw on.

The following chapters will detail the main research. The research question is stated, and research methodology discussed, followed by the research investigation and how this was undertaken. Next the four case studies will be examined followed by an exploration of key themes on the subjective experience of PCA that emerged from my discussions with the participants. How the findings relate to the literature is examined. Next a Good Practice Guide for people with PCA is proposed. Lastly, I consider if the aims and objectives of the study have been achieved and study limitations and ideas for further research and good practice development are suggested.

3.0 Chapter Three: Research Methodology

3.1 Introduction

As stated in [Section 2.6.2](#), PCA was first described in 1988 by Benson (Benson, Davis, & Snyder, 1988), who referred to it as the '*visual variant*' of Alzheimer although a small number of other conditions can, in their early stages, present with similar symptoms. These include: dementia with Lewy bodies; corticobasal degeneration; and prion diseases such as Creutzfeldt-Jakob disease (Alzheimer's Research UK/Rare Dementia Support, 2017; Schott & Crutch, 2019). Unlike tAD, people with PCA retained their memory, intellectual skills and understanding until much later in the disease (Crutch, Walton, Carton, & Shakespeare, 2017) and this allows them to clearly describe the difficulties they are experiencing. Diagnosis of PCA is often protracted and to date, the specific needs of such individuals have tended to be overlooked.

Although PCA has been recognised for more than three decades, until recently there was a paucity of research into the condition. Studies regarding the biomedical characteristics and consensus classification of this condition are being undertaken worldwide (Crutch, *et al.*, 2017) although qualitative studies to explore how people cope with living with PCA are minimal. This study recognises that exploration into the day to day challenges faced by this population is timely.

This chapter will discuss the research methodology used for the study.

The complexity of engagement with the social and physical environment for people with PCA is not well understood. This research is being undertaken to better understand the subjective experience of living with this condition and determine if the development of a sensory care model might improve the support given to this population. For the purposes of this thesis, the main interaction of interest is that of vision.

3.2 Research question

The **research question** is: “What are the specific everyday social and environmental challenges faced by community dwelling people living with posterior cortical atrophy (PCA)?”

The **hypothesis** for the study is that people with PCA have different challenges from individuals with tAD and may benefit from a specific sensory dementia care model which takes account of their visual and perceptual difficulties.

3.3 Aims and objectives of the study

The **overall aim** of the study is to explore and understand the everyday experiences of community-dwelling people living with PCA, and their caregivers. This investigation is a necessary precursor to further study around the development of a sensory model of dementia care.

Table 9 states the **objectives** of the study are to:

Table 9: Objectives of the Study

1.	Gain a subjective understanding of the everyday challenges faced by individuals living with PCA
2.	Explore the specific visual needs of community dwelling people living with PCA.
3.	Understand the stressors experienced by family members whilst caring for a person living with PCA.
4.	Identify triggers that could result in the deterioration of the individual wellbeing of people living with PCA.
5.	Consider if the development of a sensory dementia care model would benefit people living in the community who are experiencing dementia and additional sensory problems.

3.4 Management of the study

The research is being undertaken as part of a PHD conducted by myself, a student at University College London under the direction of Professor Nick Tyler, Chadwick Professor of Civil Engineering. The research is part of a

larger study funded by Alzheimer's Research UK/Economic and Social Research Council/National Institute for Health Research and based at University College London (UCL) with involvement from UCL and Brunel University.

3.5 Study Design

3.5.1 Introduction

To explore practical 'real-life' problems and issues, I decided to conduct a qualitative investigation using an instrumental multi-case study design. In-depth interviews with four community dwelling individuals living with PCA and their caregivers are undertaken. A person-centred philosophy is emphasised to enable the voices of these individuals to inform the research and analysis. A phenomenological approach is used to gain a better understanding of the specific challenges of living with PCA and/or caring for somebody with this condition.

3.5.2 Phenomenology

Phenomenology is the study of the human experience. Its aim is to explore how a person consciously perceives their everyday life situation (often called their '*life world*'). For instance, going for a walk, eating a meal, watching TV, meeting a friend etc.

Van Manen (2016) advised that its aims are about:

"... gaining a deeper understanding of the nature or meaning of our everyday experiences ... asking what is this or that kind of experience like?"

page 9

The phenomenological approach comes from a twentieth-century school of philosophy associated with Edmund Husserl (Merriam & Tisdell, 2016). He initially developed this method (with roots in both ancient Greek thought and the humanistic tradition) to be used in both philosophy, human sciences and fine art study involving the careful examination of conscious experience in every day life and science (Husserl, 1970a/1900); Wertz, *et al.*, 2011).

Smith, Flowers & Larkin (2009) pointed out that Husserl was a philosopher and his writing was conceptual mainly being concerned with how he would carry out phenomenological inquiry on his own experience rather than analysing the experiences of other people.

Leading figures in phenomenology such as Heidegger, Merleau-Ponty and Sartre developed his work further moving ideas away from Husserl's descriptive, isolated interests to a consideration of our relationship to the world and other people; extending this philosophical work to areas where the *lived experience* is relevant such as ethics, environmental studies, or humanities as well as contributing to practical professions such as health, social care and business (Wertz, *et al.*, 2011).

A leading proponent in adapting and systematizing the use of phenomenological methods for empirically based psychological research was Amedeo Giorgi. Paying careful attention to the relationship of philosophy and psychology, he articulated key procedures for undertaking this type of research (Giorgi, 2009). Other helpful step-by-step guidance in conducting a phenomenological study have been written by other authors (see Moustakas, 1994; van Manen, 2016).

When using a phenomenological approach, it is important to pay attention to a number of specific terminology and processes when undertaking this type of research. These include:

3.5.2.1 Lifeworld/Life meaning

In his last published text Husserl (1970a) described the *lifeworld*. By this he meant the every day world into which humans exist. Van Manen (2016) added to this with the term *life meaning* to refer to the way an individual understands and experiences aspects of a situation within their world. For example: a child's first day at a new school surrounded by strangers; a woman being told by her doctor she has breast cancer; a teenager leaving home for university for the first time.

3.5.2.2 *Essences*

In this context, '*essences*' means essential elements of a phenomenon. In research terms, for example, a researcher may be interested in exploring: the essence of loneliness, the essence of being wheelchair bound, or the essence of being a grandmother. In this study, my intention will be to understand the essence of living with PCA.

3.5.2.3 *Phenomenological Reduction/Bracketing*

Another important process is *phenomenological reduction*. This describes how the researcher temporarily puts aside any past scientific knowledge or subjective pre-assumptions they may have about the phenomena being studied. This is called '*bracketing*' or *epoché*, a term borrowed by Husserl from mathematics. Moustakas (1994) emphasised that this was not about forgetting past knowledge but suspending it for a while to allow the research interview to be conducted in an unbiased, receptive manner and allow critical focus on the present experience to be maintained.

3.5.2.4 *Horizontalization*

This means the process that takes place during analysis of data. At the beginning of the analytical stage, the researcher needs to consider that all the pieces of data first have equal value before then organising them into themes. At a later stage, any statements that are irrelevant to the topic and questions are disregarded. Moustakas (1994) explained that by doing so this would:

"[leave] only the Horizons (the textural meanings and invariant constituents of the phenomenon)." Page 97

3.5.2.5 *Imaginative Variation*

This involves the researcher considering the textural meaning of all the study data from various angles and perspectives. The aim of this process is to assist the researcher in recognising underlying themes and help facilitate their thinking around the development of a structural description of the phenomenon being studied (Giorgi, 2009).

3.5.2.6 Examples of Phenomenology Research

A phenomenological approach has been used in studies to focus on intense human experiences, for example: pain, loneliness, loss, frustration.

For instance: Smith & Osborn (2007); in a study considering the psychological impact of chronic benign low back pain determined that:

“If the meaning of pain to the patient is to be fully explored ... such an intensive qualitative approach is required”

and, more germane to this study, Clare (2003) suggested that this method allowed a starting point for further investigation in to understanding awareness in early stage AD stating that:

“the present study permits a preliminary modelling of the processes that appear to be involved in becoming consciously aware of memory problems and evaluating their impact and meaning ...” page 1025

Likewise, Schumacher (2010) used this approach in her study exploring caregivers of people with AD whilst coping with the demands of raising a family and the effects on their leisure opportunities asserting that:

“Phenomenology addresses the nuances of everyday experience and shows a more complete picture of the lifeworld of caregivers.” Page 1

Whilst Nowell, Thornton, & Simpson (2011), exploring the subjective experiences of personhood in dementia, stated that they:

“used this dynamic process to gain a better understanding of a participant’s life-world and their individual experiences.”

3.5.2.7 Why I chose a phenomenological study approach

Because this type of approach emphasises the lived experiences of individuals, I believed it would assist me in understanding the way (1) people with PCA subjectively experience and make sense of their social and physical world and (2) how their caregivers cope with both supporting their loved ones, as well as dealing with their own emotions as they watch their partners deteriorate over time due to this form of dementia.

I also chose this holistic approach because it reflects my concern regarding a potential lack of understanding of the needs of this population, (in contrast to the typical dementia population), and the very real impact visual and perceptual symptoms of PCA have on daily life. Because the focus of phenomenology is on the individual, using this form of analysis will allow individual stories to be explored and evaluated.

3.5.3 Reasons for using case study design

I am using case study design for the thesis because it can generate useful understanding about a particular issue or concept (Stake, 1995). The definition of case study stresses singularity and in-depth inquiry (Denscombe, 2014; Thomas, 2016). I have chosen a multi-case exploratory approach to allow me to focus on four dyadic relationships, looking at each person and each relationship in-depth. Although this small-scale study will not allow me to generalise about the lives of the overall PCA population, this approach should create a rich picture of the subjective experience of a few individuals with the condition and allow analytical insights regarding the impact of living with PCA to take place.

3.6 Breakdown of research activities

3.6.1 Ethical considerations

The study has been reviewed, and permission to undertake the research, granted by the London-Queen Square Research Ethics Committee.

The dignity, rights, safety and wellbeing of participants taking part in the study are of primary concern during the research process. Guidelines suggested by (Wilkinson, 2002) regarding issues of consent in research with people with dementia will be used. This means that informed consent will be viewed as a process of checking and rechecking with each participant that they are happy to take part and that they can refuse or withdraw at any time before or during their interview (Cantley, Woodhouse, & Smith, 2005). Care will be taken to make certain that all individuals' rights are protected and agreement to take part in the study documented.

3.6.2 Sampling

I used questions proposed by Miles and Huberman (1994) when considering which qualitative sampling strategy plan to use for this study. (See [Table 10](#)). This assisted me in deciding to use purposive sampling in order to identify participants for the study. A purposive sampling approach uses a deliberate non-random method of sampling, which aims to identify a group of people with specific characteristics (Kumar, 1996; Denscombe, 2014), in this case, the condition of PCA. I believe this sampling method will enable me to speak with individuals who are knowledgeable about living with the condition of PCA and are willing to share their experiences with me and achieve the objectives of the study.

Table 10: Checklist when considering a qualitative sampling plan

1.	Is the sampling relevant to your conceptual frame and research questions
2.	Will the phenomena you are interested in appear?
3.	Can believable descriptions and explanations be produced, ones that are true to real life?
4.	Is the sampling plan feasible, in terms of time, money, access to people and your own work style?
5.	Is the sampling plan ethical, in terms of such issues as informed consent, potential benefits and risks, and the relationship with informants?
	Adapted from (Miles & Huberman, 1994) page 34

3.6.3 Criteria for Inclusion

This is a dyadic study. Therefore, the inclusion criteria are that participants are community dwelling couples. One of each pair has a clinical diagnosis of PCA due to AD and their partner is their main caregiver.

3.6.4 Gatekeeper

The PCA Support Group co-ordinator, located at Dementia Research Centre, UCL Institute of Neurology acted as gatekeeper to identify possible

participants for the study. Once individuals agreed to be contacted, the recruitment process began with an initial telephone call from myself to the participants advising them about the project and asking if they would be interested in taking part in in-depth interviews. Four couples agreed to participate.

3.6.5 Provision of information

All participants were given an information sheet about the project in order to fully understand the aims and objectives of the study (See [Appendix Three](#)). Because the people with PCA had lost the ability to read, this information was read to them and an offer of a taped version of the sheet given. Participants were able to ask questions about the process and be reassured about the confidentiality of the study.

3.6.6 Consent

Consent forms were issued to all participants to sign (see [Appendix Three](#)) before each interview. In the case of people with PCA, only one person was able to write their signature. Two put their mark on the form and one who was unable to do this, allowed their caregiver to sign on their behalf. I conducted the study based on '*process consent*' (Cantley, Woodhouse, & Smith, 2005). This procedure entails continual explanation about the nature and purpose of the study. Therefore, at the start and throughout the interviews, consent to proceed is sought and participants reassured they can stop the interview at any time without giving a reason. To ensure anonymity, pseudonyms for participants, and other people or places implicated in their accounts are used.

3.6.7 Developing the topic guide

A preliminary brainstorming session took place with myself and Professor Tyler. The aim of the session was to review the purpose of the study, agree appropriate participants and generate ideas for questions. Ideas were also sought from Professor Crutch and members of the PCA support group. Following these sessions, an iterative process was used to develop a coherent and comprehensive, accessible framework for the interviews.

This framework was then incorporated into an interview schedule and topic guide (see [Appendix Four](#)). The topic guide was designed to allow an exploration of living with PCA in general; and how the resulting visual and perceptual problems impacted on participants' lives, including their activities, interests, relationships and identities. [Table 11](#) describes the process used to develop the topic guide used in the interviews.

Table 11: Process used to develop the Topic Guide

<p>Step 1</p>	<p>Brainstorming:</p> <p>Two sessions were scheduled. The first was with myself and Professor Tyler and the second with Professor Crutch and members of the PCA Support Group. We discussed the purpose of the study and the sort of information I would need to fulfil its objectives. These meetings generated a great many ideas for questions.</p>
<p>Step 2</p>	<p>Refining the questions:</p> <p>I then examined the list of suggested ideas and began to pull together questions ensuring that they were open-ended and key to the study. The reason why I wanted them to be open-ended was because I didn't want 'yes' or 'no' answers but rather encourage the interviewee to open up and talk about how they felt about their situation.</p>
<p>Step 3</p>	<p>Sequencing the questions:</p> <p>Questions were arranged so as to go from the general to the specific. First of all there was a series of questions to allowed them to tell the story of their life, who they were, where they went to school, their career etc before moving on to life with PCA. The aim of this route was to help they relax into the interview and come to talking about PCA in their own time rather than immediately talking about their current situation.</p>
<p>Step 4</p>	<p>Estimating time of interview:</p> <p>The interviews were timed to take from one hour to one and a half hours but I needed to be flexible with participants in case they became weary or distressed in any way. I decided I would also give them the option of two interviews if they became fatigued. My aim was that the interview should feel like a conversation rather than just questions and answers. I wanted participants to have time to laugh or to cry and to go at their own pace.</p>

<p>Step 5</p>	<p>Getting feedback from others:</p> <p>Once the first draft of questions were completed, these were sent to the brainstorming team for their feedback. Questions asked were:</p> <ul style="list-style-type: none"> • <i>Are these the right questions? Will they enable me to get the type of information I need to fulfil the objectives of the study?</i> • <i>Where the questions clear? Are any of them confusing?</i> • <i>Do the questions seem to flow from one topic to another?</i> • <i>What have I missed?</i> <p>The team were given one week to respond. Corrections were then made and sent to them again. Once they were agreed the topic guide was ready for testing.</p>
<p>Step 6</p>	<p>Testing the questions:</p> <p>The topic guide was then tested on one person with PCA and two caregivers who were not involved in the final interviews. They were asked if any of the questions made them feel awkward or were confusing. They all felt that the topic guide was comprehensive and said they felt relaxed and chatty during the process of interviewing.</p> <p>The testing of the topic guide was also useful for myself as an interviewer as it helped me learn the sequence of the questions and therefore look at the person I was talking with properly rather than reading the script. It also clarified the timing of the interviews was one to one and half hours depending on how much the interviewees wanted to share. Another key element of the testing was rehearsing how to epoché/bracketing any previous knowledge regarding PCA so I could undertake the interview with minimum presumptions about how I thought they were experiencing PCA and focus intently on their experiences.</p>
<p>Step 7</p>	<p>Signing off the Topic Guide:</p> <p>Once the feedback from the pilot session were received, the topic guide was ‘signed off’ and dates for interviews with study participants finalised.</p> <p>NOTE: The process of writing and editing the Topic Guide as well as piloting the questions took time. It is important to add at least two months into the study schedule to allow time for this to be completed.</p>
<p>Adapted from (Krueger & Casey, 2000)</p>	

3.6.8 Data collection

In total, eight interviews were conducted with four pairs of individuals: one with PCA and the other their primary family caregiver. The set up for each interview varied slightly to ensure the participant felt comfortable and reassured within their surroundings. Seven interviews took place in the participants' own homes. One caregiver asked to speak with me in a local hotel which he felt would allow him to speak more freely about his experiences.

Most interviews were conducted with individuals of each pair separately to enable them to talk in confidence about their feelings and relationship issues. One interview was undertaken with both parties present because the individual with PCA needed the support of her caregiver due to the progression of her dementia symptoms (See *Laura and David Case Study Section 4.5*). Each interview took between 45 and 60 minutes. The conversations were tape recorded and then fully transcribed.

Table 12 outlines my process for undertaking of the interviews

Table 12: Description of the process used when undertaking participants interviews

Description of the process used when undertaking participants' interviews	
1.	I first met each couple together. We had coffee and chatted about general matters such as the weather and how busy the traffic was when I was coming to meet them. This was to relax both the interviewees and myself and enable a good rapport between us.
2.	I then explained the reason for the study. (I had already done this initially by telephone but I felt that it was important to revisit the study objectives and ensure that they still felt comfortable taking part. Because (in all cases) the person with PCA had lost the ability to read, I read aloud the information sheet (see Appendix 5) and allowed them to ask questions. I then asked if they were still willing to take part. (All agreed). I also asked for their permission to tape record each session. Once they had affirmed they were happy to go ahead, I read out the consent form and

	asked them to sign their agreement. The people with PCA were unable to write but all be one made their mark. The caregivers had a separate consent form and they all signed their agreement to take part. One participant who was unable to write at all gave her verbal consent and gave permission for her caregiver to sign the form on her behalf.
3.	Interviews were undertaken singly. First the person with PCA and then their caregiver. There was one exception. A participant with PCA wanted her caregiver there for reassurance. All but one interviews were conducted in participants' homes on the same day. One interview with a caregiver took place in a local hotel on a separate occasion.
4	Before each interview I tried to ' <i>bracket</i> ' any pre-assumptions I had about PCA. Each interview began with general questions about the person's past life experiences (their career, family, hobbies etc) before turning to the subject of PCA.
5.	I tried to make the interviews a relaxed affair aiming for them to be a conversation rather than a question and answer session. This meant that the interviewee often sometimes talked around the question and expressed related memories, feelings and emotions. Although this made the interviews a little longer, this method ensured that the interviewee remained relaxed throughout.
6.	I noted interviewees tended to become emotional about their situation. I gave them time to voice these feelings and become tearful if they felt so inclined. They were all asked at points throughout the interview, particularly when they got emotional, if they were happy to continue or stop the conversation.
7.	I felt it was important not to end abruptly. Following each interview I spent time with every individual again talking about general matters until they were happy to end the conversation.
8.	The interviews lasted between 45 minutes to one hour plus extra time at the beginning and end.
9.	Following all interviews I wrote personal field notes to remind me of what had happened during the meeting and my feelings about the session.

3.6.9 Analysis

NVIVO Pro 11 (Looney , 2016; QSR International, 2017) was used during the analysis process. This is a computer assisted qualitative data analysis

3.6.9.1 *The process I used to analyse the data*

To explore study participants' experiences of living with PCA, each interview was analysed using a phenomenological approach which allowed an iterative and inductive cycle of the data suggested by Smith, Flowers, & Michael (2009) to take place. This begins with analysing the first case in detail, moving to the second case and repeating the procedure, and so on until all the case studies have been studied and recurrent themes are revealed.

Table 13 documents the analytical process used in this study:

Table 13: Analytical Process used for the study

<p>Step One</p>	<p>Reading and re-reading the data</p> <p>The narrative from the interview was read and re-read on several occasions. I also listened to the interview audio at the same time. I found that listening with headphones helped to focus my attention. This continual reading of the script as well as listening to the voice of the interviewee drew my memory back to the original interview and the rapport with the interviewee which grew as the conversation developed and helped me to become familiar with the transcript.</p>
<p>Step Two</p>	<p>Initial Note Taking</p> <p>This was a lengthy process. I started writing initial notes as soon as I started reading the transcript and added to these upon subsequent reading when I was able to begin to identify the specific ways the interviewee talked about their lifeworld. This process enable me to be aware of their feelings about how they dealt with PCA on a daily basis. I listened for key words or phrases used including important events or experiences, how they described the phenomenon; their emotions such as laughter or tears or any pauses or repetitions made during the interview.</p>
<p>Step Three</p>	<p>Drawing together emergent themes</p> <p>The continual reading of the scripts, listening to the voice of the interviewee and taking notes allowed me to create a '<i>broad brush</i>' organisational structure for my analysis using NVIVO's '<i>nodes</i>'. Nodes are discrete units of information (i.e. a passage of text or extract of</p>

	<p>audio) and I organised these hierarchically. For example, for this study as I read and listened to what the interviewee had said about the subject of <i>PCA</i>, this node began to be divided into 21 related nodes and following further appraisal of the interviews, the data was then further subdivided into other nodes, for instance the node '<i>Activities of daily living</i>' was subdivided into a further 21 nodes and some of these (for example: <i>Independence</i>) were once again divided into further nodes. This allowed me to start to create a picture of the interviewee's subjective experiences of living with PCA.</p>
Step Four	<p>Moving on to the next case</p> <p>This process was then repeated for all the interviews.</p>
Step Five	<p>Looking for patterns across all interviews</p> <p>Once all the cases had been reviewed and I had time to reflect on the notes I had taken. I started to pull similar information from the interviews together. Appendix Five shows a snapshot of the nodes I created from information gleaned from the interviews and how many times each subject was mentioned by each interviewee</p>
Step Six	<p>Resulting Themes</p> <p>The analysis was then mapped into the seven main themes which appeared to be the most important experiences of the interviewees. Table 14 below outlines the themes:</p>
<p>Based on suggestions by (Smith, Flowers, & Michael, 2009) pages 80-99</p>	

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Table 14: Main Themes from Analysis of Participant Interviews

Main Themes from Analysis of Participant Interviews	
1	Awareness and Understanding of PCA
2	Journey to Diagnosis
3	Losses
4	Coping
5	Social Supports
6	Facing the Future
7	The emotional impact of living with PCA

A comprehensive commentary of the themes can be found in [Chapter Five: Findings: Themes.](#)

3.6.10 Security of data

To protect the anonymity of individuals, all participants, places and people named by interviewees have been given pseudonyms. Electronic information was stored in a password protected file. Paper copies of transcripts were kept in a locked cabinet and were destroyed at the completion of the study.

3.6.11 Work Schedule

Initially, the research was due to be undertaken over a 12-month period. However, for personal reasons, the schedule was extended by a further 12 months to allow completion of the study.

3.6.12 The Step by Step Process of this research study

Table 15 below sets out the step-by-step process of how I undertook the research with additional comments which may be helpful to any reader who wishes to replicate this type of study.

Table 15: Step by Step Process of how I undertook this research study

Step-by-Step process of how I undertook this research study		
No	Activity	Comment
1.	Choosing the topic	Because this was a PhD study I knew I would need to spend many hours exploring the subject. It was therefore vitally important that the subject was something in which I was extremely interested. Vision loss and dementia were key aspects of my work and I was keen to learn as much about the subject as possible. A meeting with the PCA support group in London alerted me to the paucity of subjective research in this area and this was why I chose the topic.
2.	Background Reading	The main bulk of the background reading was done in the first year but in reality it carried on throughout the project to ensure I was aware of any new published research.
3.	Deciding research type	Because I was exploring subjective experience of people with PCA, I decided that the study should be qualitative. Phenomenology was chosen because of its emphasis on a person's lived experience. Case studies were chosen because they would involve a comprehensive exploration of individual experiences.
4.	Choosing type of Sampling	Purposive sampling was chosen because PCA is a rare atypical dementia and numbers of individuals diagnosed with this condition are relatively small. It would therefore be difficult to find people with this problem amongst the typical dementia community.
5.	Finding gatekeepers	The co-ordinator of the PCA support group was the gatekeeper who found participants. This was because she had a large database of group members and was willing to make contact with people through the PCA newsletter and by phone.

6.	Deciding on the number of participants	I decided to have a total of four couples – one with PCA and one the caregiver. I realised I would need to interview and transcribe all the data myself and this number seemed the most manageable and would allow me to conduct an in-depth study of each person in the timeframe of the project.
7.	Writing the topic guide/ information sheet/consent forms	<p>The topic guide/information sheet was completed in 6 stages:</p> <ol style="list-style-type: none"> 1. Two brainstorming sessions 2. Initial draft written by me from topic ideas suggested at the brainstorming meetings 3. First draft was sent to all participants of the brainstorming sessions and asked to reply via email. 4. Second draft then written up and sent out again for approval. 5. After approval, the topic guide/information pack were piloted with three people who were not involved in the final research. Two caregivers and one person with PCA. They were asked if the questions made sense or if they needed refining. It also enabled me to practice ‘bracketing’. 6. Topic guide was then finalised.
8.	Ethical Approval	The study was reviewed, and permission to undertake the research, granted by the London-Queen Square Research Ethics Committee.
9.	Finding Participants	People who were contacted by the PCA support group co-ordinator were asked if their details could be sent to me. Once they had agreed, their telephone numbers were passed to me and I contacted each couple. I explained about the study and asked if they were willing to take part.
10.	Arranging interviews	Interviews were arranged via the telephone

11.	How the interviews were undertaken	<ol style="list-style-type: none"> 1. I first met each couple together. We had coffee and chatted about general matters. This was to relax both the interviewees and myself and enable a good rapport between us 2. I explained the reason for the study. Because (in all cases) the person with PCA had lost the ability to read, I read aloud the information sheet and allowed them to ask questions. Once they had affirmed they were happy to go ahead, I read out the consent form and asked them to sign their agreement. The people with PCA were unable to write but all but one made their mark. The caregivers had a separate consent form and they all signed their agreement. The participant who was unable to write gave her verbal consent and gave permission for her caregiver to sign the form on her behalf. 3. Interviews were undertaken singly. First the person with PCA and then their caregiver. 4. Before each interview I tried to 'bracket' any pre-assumptions I had about PCA and how it would affect the participants. This needs practice so rehearse doing this before the interviews. 5. I tried to make the interviews a relaxed affair aiming for them to be a conversation rather than a question and answer session. 6. Following each interview I spent time with every individual again talking about general matters until they were happy to end the conversation. 7. The interviews lasted between 45 minutes to one hour plus extra time at the beginning and end.
12.	Taking Field Notes	Following all interviews I wrote personal field notes to remind me of what had happened during the meeting and my feelings about the session.
13.	Computerised Analysis Tool	There are a number of software packages on the market. In my case analysis was undertaken using NVIVO Pro 11. This is a computer assisted qualitative data analysis software tool developed to aid researchers analyse semi structured interviews, field notes, journal articles and web pages. It allowed me to create a dataset of all the different types of information I collected for the study.

		(It is important to understand how to use the software for inputting and analysis so study time is not wasted. This was the first time I had undertaken analysis using an computerised tool so I undertook online training before starting the process)
14.	Analysis	<p>A process of six stages were used in the analysis process:</p> <ol style="list-style-type: none"> 1. Reading and re-reading the transcripts 2. Initial note-taking 3. Drawing together emergent themes 4. Moving on to the next case 5. Looking for patterns across all interviews 6. Resulting themes
	Anonymity	To protect the anonymity of individuals, all participants, places and people named by interviewees have been given pseudonyms. Electronic information was stored in a password protected file. Paper copies of transcripts were kept in a locked cabinet and were destroyed at the completion of the study
16.	Writing up	The study was written using MS word, PowerPoint and excel. Initially, the write up was due to be undertaken over a 12-month period. However, for personal reasons, the schedule was extended by a further 12 months to allow completion of the study.

4.0 Chapter Four: The Case Studies

4.1 Introduction

This chapter describes four dyadic case studies where one person has PCA and their partner is their main caregiver. From semi-structured interviews between each couple and myself, the story of their continuing journey of living with PCA emerges. Most individuals were interviewed alone to maintain confidentiality and encourage people to be as open and honest as possible. In [Case Four](#), the participant with PCA was experiencing significant dementia symptoms and so the couple were interviewed together, although a confidential interview was also held with the caregiver separately. The names of the contributors and any place names are changed to preserve anonymity.

The interviews are structured around a topic guide which reminded me to ask the same pertinent questions to everyone but allowed for the conversation to flow freely to encourage participants to relate their experiences as comprehensively as possible without feeling pressured.

Following analysis of the interviews, each couple's story is told by combining their separate interviews. In this way it is possible to compare and contrast their responses to the same questions.

4.2 Case Study One: Betty and Harry's Story

Betty and Harry were interviewed at their home in separate rooms to enable complete privacy. Each interview took one hour, was tape recorded and fully transcribed.

4.2.1 Setting the Scene

Betty is a 74-year-old lady who had been diagnosed with PCA two years ago. She appears a very sociable and chatty person. Betty has been married to **Harry** for 52 years and has three grown-up children and one grandchild. She and Harry have lived in **Chester** all their lives except when Betty attended teacher training college when she was eighteen. Apart from short breaks when having her children, Betty was employed as a teacher her entire working life finally retiring at 60 years. Although working full time, Betty was a real home-maker particularly enjoying cooking and entertaining. The activities she most appreciated in her spare time were reading, painting, embroidery, gardening and walking in the countryside with a group of lifelong friends. She also helped with flower arranging at her local church. During her younger life her general health was good although she had suffered from mild eczema from childhood. She had had a cataract removed in her 60s and now wears spectacles for close work. Harry is 77 years old. He left school at 15 years and undertook an apprenticeship in gaining a national certificate in engineering. However, disappointed with his job prospects he changed direction and became a salesman. For the last ten years of his working life, he worked for himself, finally retiring at 70 years. Harry has diabetes, which is under control, and an essential tremor in his left hand.

After living in a large detached house for many years, the couple down-sized to a smaller property three years ago and now live in a ground floor flat in a quiet area of town fairly near their former home. The premises consist of two large bedrooms, one ensuite containing a shower and toilet. There is a kitchen, separate bathroom and large dining room/lounge leading through French windows to a sunny patio sitting area. The flat has good natural lighting. The colour scheme on the walls is cream with plain

beige carpets. The kitchen and bathroom areas have lino floors in plain design. The bathroom suite is white with little contrast. There are no hand rails to assist people to rise from the bath or toilet. The kitchen is very modern containing a washer/drier, dishwasher, microwave, fridge/freezer and cooker plus a dark wooden kitchen table and chairs. The kitchen cabinet doors also have a wooden finish. The dining room/lounge contains a bright red sofa and matching armchairs and a large dining table and chairs. The flat is extremely tidy with flowers and plants decorating the main areas.

4.2.2 Realising something was wrong

Betty describes her working life as very enjoyable and she was fit and well. However, as she neared retirement she felt sometimes she made mistakes or felt clumsy doing things. Harry says looking back he also had noticed little incidents that didn't seem quite right.

<p><i>“I was alright all my working life it was quite normal. It was only just little by little things were going awry (Betty)</i></p>	<p><i>Identifier: PCA1.1</i></p>
<p><i>“Long time ago ... Oh now, 8 years, probably more ... things used to happen. I used to say to her ‘what have you done? Why did you do that? (Harry)</i></p>	<p><i>Identifier: C1.1</i></p>

Comment: *Betty and Harry appear to be describing the very early stages of dementia with minor incidences being thought of as just the process of getting older.*

Then in 2004, just after retiring, Betty noticed something was not right with her vision when she began to experience difficulties in driving, particularly not being able to judge how close she was to other road traffic users.

<i>“I felt that I was going to hit a car that was alongside me ... I was pulling my arms into my body as if I could squeeze the car.” (Betty)</i>	Identifier: PCA1.2
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Comment: Betty seems aware that ‘pulling her arms in’ would not resolve the problem of her perception of the relative positions of the two cars.

Harry advises he was also noticing changes in his wife’s visual abilities.

<i>“... there were so many little things... a level of something that I found different. We’d been married a long, long time and you get to know people very well indeed and to me it was obvious but to her it wasn’t because she was the victim to whatever it was.” (Harry)</i>	Identifier: C1.2
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After bumping the garage door, Betty was concerned about her vision so went to see her optician for a routine appointment.

The optician didn’t flag up any serious problems and so she carried on driving for another two or three years. However, she then began to have trouble in driving at night.

<i>“I think it was the change from daylight to dark, and if you are a bit wary already it’s even worse. I think that’s when I thought I am not going to go out in the dark in the car.” (Betty)</i>	Identifier: PCA1.3
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Harry also noted that her driving had become erratic.

<i>“She was always a brilliant driver. Very careful. I had every confidence in her ability to drive. But then she (pause) ‘oh I would have let that chap out, you know’ and ‘oh, you’ve got an amber light here.’ Just silly things.” (Harry)</i>	Identifier: C1.3
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They were so worried they both went to see her GP but was told that it was probably stress and Betty was prescribed sedatives.

<i>“We went to the GP and he said, ‘oh sedatives, it could be stress,” (Harry)</i>	Identifier: C1.4
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Betty describes that she felt her vision getting worse particularly judging how close she was driving next to other vehicles but because both the GP and the Optician had not flagged up any specific problems with her eyes, she continued to drive her car during day time hours.

<i>“I found that I, I felt that I was going to hit a car that was alongside me. I couldn’t judge how close I was to things. ... but I carried on driving for ... two or three years, I think.” (Betty)</i>	Identifier: PCA1.4
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Comment: *The Alzheimer Society (Alzheimer's Society, 2016) have reported that one in every three people with dementia still drive. It could be argued that Betty’s visual/spatial problems are limiting her ability to drive safely but at this point, Betty doesn’t have a diagnosis of dementia or PCA and the optician and GP haven’t determined any serious health issues, so she continues to drive for a number of years even though she knows her vision is getting worse.*

4.2.3 Journey to diagnosis

After a while, Betty’s vision and perceptual problems seemed to be getting worse and so she and Harry went again to the GP and asked for a second opinion.

<i>“It could be this, it could be that. I wasn’t interested in what it could be, I was interested in what it was. So, after a while, quite a while, I thought, I have to fire this up and could we see someone”. (Harry)</i>	Identifier: C1.5
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In 2011, Betty was sent to the local general hospital and underwent a scan and was then referred to a specialist psychiatrist where she was given a battery of memory and written tests.

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<i>“I had a scan, but it didn’t show up anything for some time” (Betty)</i>	Identifier: PCA1.5
<i>“He said, well your memory is fine. It’s better than most people. I thought that’s good but we’re not getting anywhere. I was going every three or four months to check if I had got better or worse and doing other tests. I had to build things, put things straight, draw something. Well it wasn’t too bad at first but then that deteriorated. I couldn’t remember how to join things up!” (Betty)</i>	Identifier: PCA1.6

Betty was told she had visual agnosia and continued to be monitored at the hospital. Harry expressed feelings of concern over what the condition actually was.

<i>“We were concerned about the word ‘dementia’. Is it a form of dementia? It’s visual agnosia”. (Harry)</i>	Identifier: C1.6
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Finally, after four years she had another scan and was finally diagnosed with PCA in 2015.

<i>“I think it did go on a long time but you see all the time I was being monitored but they, I suppose, this being a rare thing, it wasn’t considered, I suppose, I mean I just felt well I am going back and forth, back and forth, and until something drastic happens I think or you get absolutely fed up and you think well there must be something wrong and that was when it was diagnosed. ... When they told me, I’d never heard of it.” (Betty)</i>	Identifier: PCA1.7
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When I ask what they understand PCA to be, both know it as a problem with brain function rather than eyesight, but explain it differently:

<i>“It’s not my eyes, it is exchange of ... what I am seeing into where it should be. It’s just blanked off” (Betty)</i>	Identifier: PCA1.8
<i>“We’ve been more or less told that it was not a visual problem, it was a perception problem. The message is going through, but it is not going any further. It is hitting a wall.” (Harry)</i>	Identifier: C1.7

Dementia medication has not been offered to Betty. She takes a statin, antidepressant and soluble aspirin daily.

<i>“I’ve got these happy tablets (laughs). I don’t really think they make any difference to be honest, but I take them and think ‘Oh well they can’t do any harm’” (Betty)</i>	Identifier: PCA1.9
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Comment: *Betty has now been struggling with her vision for some time and it is only the insistence of the couple that the GP referred Betty to the hospital. At this stage her memory is good so there are delays in getting a proper diagnosis with many trips to see various clinicians at the hospital. Once diagnosis is made, they both appear to have had a good explanation of the symptoms as they can understand and describe the condition of PCA.*

4.2.4 Reactions to knowing

The couple responded pragmatically to diagnosis. For Harry, who acknowledged that his wife has a progressive rare form of Alzheimer’s, at least understands what is happening to Betty and wants to try and manage the condition as a team.

<i>“I said to the consultant when it was revealed, I said, it hasn’t happened to Betty, it’s happened to us. So, it’s a situation that is there to be managed. As it develops we’ll manage it differently, but it is important to manage ourselves as well and take whatever guidance or assistance may be offered to keep our own sound partner at maximum operating efficiency. I was a great one for food, pork pies but not now. So, I went down to the docs and had a well man check and every year now because I’ve got to look after Betty. (Harry)</i>	Identifier: C1.8
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Betty appears quite down-to-earth about her diagnosis taking great comfort from Harry’s assurances of support.

<i>“Well to be honest I didn’t weep, no, no. I thought well at least we know what we’re about. Fortunately, Harry is very supportive. He said ‘Right there are two of us in this. We’ve just got to get on with it and keep as normal as we can’. Well, I felt comforted. I mean after 50 years together you know each other back to front. He said things like ‘I’ll never leave you. ... I can take care of you’.” (Betty)</i>	Identifier: PCA1.10
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Comment: They both appear to take the diagnosis in a practical rather than emotional way at this point. Harry is determined for them to work as a team to manage the condition. Betty seems comforted by Harry’s reassuring words.

4.2.5 Day to Day challenges

This next section looks at the challenges faced daily by the couple as they manage Betty’s progressive condition.

4.2.5.1 Personal Care

Although Betty can go to the toilet by herself, other personal care problems arise daily. Harry thought Betty was happy to shower or bathe herself, but Betty describes a fear of falling in the bath:

<i>“Well, I’m alright getting in, but that bath is very deep. It’s a big bath. Getting out, it’s because there are no hand rails. I wish we had one but those things you put on the wall, I don’t trust them. I think it would have to be a real big job and hammered on. ... At the moment, I am capable of going on my hands and knees and then I can pull myself out. I don’t know. I think I might fall.” (Betty)</i>	Identifier: PCA1.11
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Harry advises that Betty finds difficulty in distinguishing the washing products but pushes her to succeed on her own.

<i>“I say, ‘Ok which one do you want?’ There’s silence. ‘Come on touch it, touch it, pick it up. Was it this one?’ I say. ‘you’re deciding’ cause I’m not soft with her.” (Harry)</i>	Identifier: C1.9
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Comment: Betty demonstrates a fear of falling and there are no adaptations in the bathroom to help her. A hand rail would be of benefit. The council's occupational health team may help here. It appears she is not able to differentiate between washing products and Harry is encouraging her to make her use her other senses to choose products. This difficulty may be due to being unable to read the labels of the products (*alexia*) or the inability to identify objects (*associative agnosia*).

4.2.5.2 Dressing

Dressing is also difficult. Betty explains her problem:

<i>"It's getting things the right way up. I don't know whether that's the top and I sort of put my arm down the thing, the hole. It's assembling the garment. Then say it's a pair of knickers. I pick them up and I think that's the back and then whether I twist not knowingly but then it goes wrong. So, then I turn them inside out, upside down and in the end, I yell for help. ... I mean I've got to the point it's awful to get dressed'."</i> (Betty)	Identifier: PCA1.12
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Harry muses wryly:

<i>"normally I have to help her get the bra the right way instead of inside out or back to front. I said, 'my god, men are so lucky they don't have to manage with one of these harnesses every day.'" (Harry)</i>	Identifier: C1.10
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Betty expresses great frustration when it comes to dressing because of the difficulties she is experiencing.

4.2.5.3 Food preparation

Betty says she enjoys cooking and baking, but these tasks have now been taken over by Harry. Betty talks about needing to relearn doing things and expresses that she doesn't think the effort is worth it.

<i>"I enjoyed cooking very much. I made my own bread but sadly all these things have gone by the board. ... I can't do it now. I can't measure anything out you know flour and whatnot. ... That's a problem. ... It's just now I've got out of the habit ... I think relearning how to</i>	Identifier: PCA1.13
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<i>bake a cake ... is it worth the effort when you can buy one” (Betty)</i>	
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Harry recalls enjoying Betty’s baking in the past but explains another reason for her problems in the kitchen and the possible dangers:

<i>“She doesn’t do any cooking or baking. None whatsoever. She was a terrific cook. She did loads of it. Wonderful cook. ... [Looking at the cooker] but she can’t perceive what she’s looking at. Looking is not understanding. She says: ‘I’m used to gas’. ... Now that causes me concern because it wouldn’t be difficult to get a gas top but then we would have living flames, tea towels, no, no”. (Harry)</i>	Identifier: C1.11
<i>“If I would say ‘can you bring the butter dish across’ ... this is what would happen. [goes around the room touching and feeling]. ... She looks for things like a blind person. That’s what she does. She’s touching surfaces all the time. Waving her hands about. (Harry)</i>	Identifier: C1.12

Comment: Several PCA symptoms appear to be making it difficult for Betty to bake and cook – her deteriorating motor/perception/memory skills, her inability to read labels and recipes (*alexia*) and her numeracy skills in working out weights and measures (*acalculia*) and this is resulting in lack of confidence in herself. Harry appears extremely worried about Betty hurting herself. Could RNIB products such as talking scales, talking label covers, liquid level indicators help here?

4.2.5.4 Eating and drinking

Betty has trouble physically eating. She finds it impossible to use a knife and fork properly and prefers finger food although can still use a spoon. She also has problems seeing food on the left side of the plate. They dine out quite often with their friends at a regular restaurant who were very accommodating and between them they continue to make the occasion a normal and pleasurable experience:

<p><i>“When we book a table, we say when we come in can you send us to a corner table, so Betty can face me, and I can look out into the restaurant. So, if she wants to eat with her fingers she can do. No problem they say so they are very good.” (Harry)</i></p>	<p>Identifier: C1.13</p>
<p><i>“When we go out for meals if she orders gammon and I order something else. When they bring the food and the girl says ‘Gammon?’, I say, ‘yes down here please’ and she has mine. Then I get the steak knife that comes with the gammon and I cut it all up for her. Then I do a swap and that works well. Our friends do the same for her without saying anything.” (Harry)</i></p>	<p>Identifier: C1.14</p>

Comment: Betty appears to have *left sided visual neglect* resulting her not seeing food on the left side of the plate. *Apraxia* is affecting her ability to use a knife and fork. However, Harry and their close friends are enabling life to go on as normally as possible as they are still able to eat out and socialise.

4.2.5.5 Reading and numeracy

Betty gradually lost the ability to do many of the activities she used to enjoy. Formally a great lover of books, she is now unable to read or write her name.

<p><i>“Well it frustrates me that I can’t read properly. I am unable to write my name properly and I used to read, and it would be wonderful if I could sit down and just read. I’ve tried magnifying glasses and all the rest of it. Isn’t it awful, awful. ... but anyway, with the talking books, that’s good. I had one of them last week and I enjoyed that. (Betty)</i></p>	<p>Identifier: PCA1.14</p>
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Numbers are also a problem. Using the telephone or reading a clock are challenging. Again, the use of accessible gadgets is proving helpful. Betty explained:

<i>"To dial, numbers seem, how can I say, they move. I just can't see them. They are not jumping I just can't, I can't do it. ... I'm in the process of the boys transferring all the numbers I would use so that Number 1 is my daughter. You see even if I get a few numbers if I take my eye off to then dial the next one, it seems to have disappeared. That links with the clock. I can't read the time either. Which is why I have a talking clock. ... That's marvellous. ... You've got the date and the time and that's a great comfort. (Betty)</i>	Identifier: PCA1.15
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Another difficulty Betty has is the inability to identify money correctly or physically use a credit card or ATM machine which means that her independence is further eroded as she needs to depend on others to manage her finances. Harry explains the problem.

<i>"I wouldn't let her go with credit cards because she can't use them, and she couldn't sign her name and she couldn't do the numbers or draw money from an ATM she can't do that." (Harry)</i>	Identifier: C1.24
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Comment: Betty's *acalculia*, *agraphia* and *alexia* symptoms are causing her great distress as she cannot understand money, read or write her name any more. Fortunately, she appears to be able to carry on getting pleasure from fiction using audiobooks. Her description of numbers jumping and moving when she tries to use the telephone could be describing *visual disorientation* or *ocular motor apraxia*. The use of a simple button phone and a talking clock are helping her maintain her orientation to day and time and help her keep in contact with friends and family.

4.2.5.6 Problems with perception

Harry explains that sometimes Betty will not see him or is unaware of where he is sitting or standing.

<i>"It's not recognising, I think it's seeing. She walked out of the kitchen the other day and I had just come out of the bedroom and she was just looking ahead carrying a cup of coffee and had I not stopped her, she would have walked straight into me." (Harry)</i>	Identifier: C1.15
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<i>"I can go in the lounge when she is sitting watching television, especially on the left-hand side, where her peripheral vision is practically zero and she doesn't know I am there. No idea I am there. ... her eyes are perfectly alright. She has no problem there. She just doesn't see. ... also she brought a tea the other day and she took it over to an empty chair and said here's your tea." (Harry)</i>	Identifier: C1.16
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Betty also remembers the incident:

<i>"I remember making Harry a cup of tea and when I came back I can't quite remember the order of what I said but the fact was it looked like him, the shape. It was winter, and we didn't have the lights on and I saw this shape and talked to this saying 'don't let it get cold' and just at that moment I heard him cough and I thought I am looking at something and thought it was something else. It has happened a few times" (Betty)</i>	Identifier: PCA1.16
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Betty speaks about putting objects down and then not being able to find them again. She explains this away by saying she is probably doing too many things at once.

<i>"I put things down and sometimes I can't find them. Harry comes in and says here they are. I think my trouble is that probably I try to multi-task and I'll think I'm doing something there and the phone rings and I put that down and then I think where have I put it? Things disappear!" (Betty)</i>	Identifier: PCA1.17
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Comment: Harry doesn't mention the lighting but could the problem Betty's experiencing of not seeing him be down to [low light levels](#) or her [visual neglect problem](#)? She says this type of incident has happened several times, so she could be experiencing [simultanagnosia](#) and not perceiving the whole scene within the room correctly. When she describes putting things down and then losing them it may be due to either forgetting where they are or losing visual sight of them.

4.2.5.7 Memory and insight

Betty has been experiencing visual and perceptual problems for 14 years and has been diagnosed with PCA for two years. Nevertheless, she can understand and respond coherently to the questions asked although she

feels that sometimes her memory does not function too well during the conversation and this leads to feelings of frustration and lack of confidence.

<i>“I’m trying to think which is difficult, oh God” (Betty)</i>	Identifier: PCA1.18
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Harry feels that Betty’s memory varied but particularly her short-term memory is affected:

<i>“She can remember remarkably sometimes. Short term sometimes is faulted. It varies, varies a lot. ... Now she can start a conversation and with the preamble to the conversation, she gets to the main point and she can’t remember what the main point was. I say when that happens again come out with the main point and I’ll fill in the preamble myself.” (Harry)</i>	Identifier: C1.17
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Comment: Betty’s difficulties have been present for 14 years and though both she and Harry recognise her short-term memory is not as good as it has been, nevertheless she is still able to understand and respond well to conversation and questions. Would somebody with tAD for this length of time still be able to cognitively function as well?

4.2.5.8 Environmental Challenges

Betty explains that often her energy levels are low which does not motivate her to do any forms of exercise although Harry blames himself for using the car too much.

<i>“I am not quite sure about my energy because I don’t do enough, you know, I should do more. Get outside.” (Betty)</i>	Identifier: PCA1.19
<i>“She could do more but that’s my fault. .. well because I say, it’s winter, let’s use the car to go down to town. ... Yes, but it’s just an excuse.” (Harry)</i>	Identifier: C1.18

Betty also describes several experiences of environmental challenges both inside and outside the home.

4.2.5.8.1 Balance, walking and fear of falling

Visual-spatial difficulties cause problems for Betty every day. It affects both her balance and walking resulting in her being in danger of falling.

Betty dismisses this as a problem due to her age:

<i>“I would say I am not as steady on my feet as I was. Balance is a bit of a muddle. I can be walking and then I wobble a bit. ... I have done quite a few trips, not falling down but cobbling over, that sort of thing. But I think that is fairly common with people as they age.” (Betty)</i>	Identifier: PCA1.20
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But a bad fall at a friend’s house has knocked her confidence:

<i>“... We had a lot of steps [at our previous home]. I was used to steps. I used to go two at a time up the stairs but alas no more.” (Betty)</i>	Identifier: PCA1.21
<i>“I fell two or three years ago. I had an awful fall. I was at a friend’s house and they had a Victorian staircase. ... I went upstairs to the loo, walked down and I stepped out into nothing and I crashed over the bannister. I just fell, fell and I clung onto the thing and landed in a puddle ... oh it was really frightening.” (Betty)</i>	Identifier: PCA1.22

Harry remembered the incident but does not perceive falling as a major problem:

<i>“About four years ago she just toppled forward but I’ve not noticed it since. I’ve not noticed her falling. ... well she certainly hasn’t fallen in the house or anything like that. ... No, it’s not a problem, at present” (Harry)</i>	Identifier: C1.19
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However, the stair incident has left Betty with a fear of falling. She explains what happens when faced with such a situation.

<p><i>“Going down stairs, as long as I have a rail I feel safe, but I could no more be on a flight of stairs and walk down without holding on. I feel like I am being pulled. Pulling me down. It’s a feeling inside. I start wobbling inside. ... I get a bit panicky, yes and sometimes steps, I can’t, I panic a bit because the step’s there, but I am not sure if there is another one or I have been on one. I mean, it’s torture.” (Betty).</i></p>	<p>Identifier: PCA1.23</p>
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Comment: Betty’s remark ‘pulling me down’ is interesting. Is this because she is so frightened of falling? She uses words like ‘frightening’, ‘panicky’ and ‘torture’ to describe her feelings. Betty doesn’t seem worried that her legs will give way. She appears to be describing some sort of perceptual reaction.

Certain surfaces also prove to be a problem to walk on when Betty is out shopping and makes her wary of falling:

<p><i>“I think it just makes it a muddle. I have the sensation of ‘oh I can’t walk on that’ sometimes on certain floors in shops. Patterns and shiny floors are a problem. There’s one in a shop that looks like a glass panel. I mean I don’t know if it was glass, but it was shiny and black, and it looked like a hole you see. I stood at the top and I thought, ‘I can’t move’. A lady said, ‘are you all right?’ and I said yes, yes I am, as you do’ (laughs). And I won’t go in that shop again because I am not confident enough to trust myself to not deliberately fall” (Betty)</i></p>	<p>Identifier: PCA1.24</p>
<p><i>“Pavements are hazardous to her ... because some pavements are so uneven now. The Council is running out of money to put them right. A raised paving slab, a curb, step out curb up curb down. Another entrance to a drive, I said ‘no curb just keep going’. It just becomes second nature.” (Harry)</i></p>	<p>Identifier: C1.20</p>

Comment: Betty's describes certain surfaces as being difficult to walk on. Her deterioration in visual capacities such as a *heightened sensitivity to glare* and her *contrast sensitivity* and *depth perception difficulties* appear to be impacting on her experience of different floor surfaces.

The couple differ in their response to the question about falls. It is obviously a great worry for Betty, quite understandably as she has experienced falling down stairs, but Harry seems almost to dismiss that falls are problem due to Betty's PCA, rather that the problem is their local pavements are poorly maintained. However, as he continues the conversation it is clear he doesn't think Betty can see the walking environment correctly.

Although stairs are a challenge for Betty, she explains that she would never choose to take a lift instead.

<i>"I wouldn't take the lift because of the buttons. I'm hopeless at pressing buttons. I can't recognise which door to get in things or out of things and if I do go in a lift I make sure Harry is by my side. It just gets me. You know when you go a bit churny."</i> (Betty).	Identifier: PCA1.25
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Comment: Betty symptoms of *visual disorientation* and *ocular motor apraxia* may be preventing her from being able to work out which button to press in the lift. Once again, she displays signs of distress and panic at the thought of trying to do this without Harry's support.

Another problem Betty reports is sometimes not perceiving the environment correctly. She has fallen between the bed and wardrobe whilst staying at a hotel and is very wary when sitting down on chairs, in case she misses the chair and lands on the floor.

<i>"I was coming into a bedroom in the hotel, we'd be out walking around, and I come in and it was hot, and I flopped down on what I thought was the edge of the bed, but it wasn't, it was a narrow gap ... I fell backwards through the gap."</i> (Betty).	Identifier: PCA1.26
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In her usual pragmatic way, Betty dismisses the event:

<i>“Well as long as it’s just me there (laughs) ... it’s just a silly nuisance. There is no point in crying over spilt milk attitude. You’ve just got to think you’ve done a daft thing here. Daft’s the word” (Betty)</i>	Identifier: PCA1.27
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However, such incidences knock her confidence and further increases her fear of falling.

<i>“I am aware now that if I am going to say a restaurant I really have to look at the seat. I can’t just walk in and sit down. I mean if Harry or a friend is nearby they pull the thing out and say sit there but sometimes I have to be very cautious which makes it very slow” (Betty).</i>	Identifier: PCA1.28
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On the other hand, Harry doesn’t appear to have such concerns and feels their friends sometimes do too much to assist Betty.

<i>“We have two very dear friends who as soon as she goes into the restaurant leap up and go and help her. She doesn’t like it, she doesn’t need it. All it does is simply endorse her condition to her. ... They are lovely, but it is counterproductive.” (Harry)</i>	Identifier: C1.21
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Comment: Betty is describing [problems with depth perception](#) as well as not perceiving the environment properly ([simultanagnosia](#)) which are having a great impact on her confidence levels. However, Harry appears not to realise this, proposing that she doesn’t need or want so much help and it only serves as highlighting the fact that she has PCA.

4.2.5.8.2 Getting Lost

Getting lost and disorientated is beginning to become a real issue for Betty. This is occurring both inside and outside the home.

<i>“I repeatedly go in the wrong room. I just don’t know why it happens, but it does. As soon as I get over the threshold I think ‘oh foolish’.” (Betty).</i>	Identifier: PCA1.29
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She recalls an extremely upsetting incident:

<p><i>“...only the other morning, I was wandering around and I didn’t know where I was. I could see they were my possessions but where they were in what position they were I hadn’t a clue. I think it was partly that the room had all been changed, things were there that weren’t there the day before. ... I just got upset, really upset. It was the first time I felt ‘oh I don’t know why this has happened because I must know where I am, I must be at home’? It was very confusing. ... I was walking around feeling. Feeling on the walls. ... I just broke down. I thought this is terrible, terrible. I couldn’t see anything, so it was all feel and I was going round and round and touching things. ... I touched Harry’s foot and I thought ‘it’s a foot, it’s a foot’ so I knew it must be his. I think I do a lot by feel now. And Harry said, you’d think you were blind, and I said, ‘well in a way I am’.”</i> (Betty)</p>	<p>Identifier: PCA1.30</p>
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Harry also remembers the event:

<p><i>“I was in bed. ... she came in very very tearful. ‘I thought I’d lost you’ ... she couldn’t see me visually and something was sticking out, it was my foot and I felt her get hold of it. It was at the time when everything was moved around because we were having furniture cleaned. It was dark ... and she got hold of me and you could see relief written all over her face. Oh, she just cried, sobbed. I said, ‘come on’ and I moved over, and she got in and I said, ‘come and have a cuddle’. I said, ‘God what’s the matter with you, crackpot?’ That’s how we handle things.”</i> (Harry)</p>	<p>Identifier: C1.22</p>
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Betty is also prone to getting lost outside the home.

<p><i>“I got lost in Marks and Spencer. I’d gone in one door and I’d come out of another door and it was all foreign. I thought well I don’t know where I am, I can’t see a door. Anyway, there was a fella doing some work and I said this might sound very silly, but can you tell me how to get to Marks and Spencer? ... I have asked people because people are so kind I think well if it was round the other way I would do it for somebody. It’s no great deal really. But Harry doesn’t like me going out on my own because he worries I am going to get lost because I do get lost. (Betty)</i></p>	<p>Identifier: PCA1.31</p>
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Harry has witnessed Betty getting disorientated and expresses the need to protect her from herself by not letting her venture out alone:

<i>“I dropped her off in town to go to the hairdressers. I saw her get out of the car, cross the road and instead of going into the hairdressers she turned left and set off walking up the high street. She had no idea where she was going. I wouldn’t let her go to town on her own because she has this happy look about her like there’s nothing wrong. ... It’s best when the first suggestion that there are difficulties they’re to be stopped stone dead because it is not going to get better, it’s going to get worse” (Harry)</i>	Identifier: C1.23
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For Betty, the loss of being able to go out shopping by herself means losing one of her greatest pleasures in life. She explains it is not only the enjoyment of shopping but the independence of doing something by herself when she wants to do it:

<i>“... I used to go by myself shopping and that has been taken away from me and I did really, I won’t say it was a hobby, I’m just like most women, I like to browse around. You can’t browse around with a man (laughs). ... I went out with a friend and ...it was lovely cause she is very bouncy and she’s encouraging, and I really enjoyed that. But you don’t want to ask people permanently, you know. I suppose it’s because I’m frustrated that I’m not independent, you know if I want to go out, I used to go out. Now I have to wait. I have friends and I know they would go but it’s a fact that I like to go by myself”. (Betty)</i>	Identifier: PCA1.32
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Comment: Betty is experiencing *environmental agnosia* causing her to get lost both outdoors and in her own home. Harry explains he will not let her go out on her own because he is convinced she will get lost. This has meant Betty has lost a major part of her independence and freedom to go out when and where she wants but she is very aware that she is likely to lose her way and so appears to be resigned to the situation.

4.2.5.8.3 Light and Dark

Betty is cautious about wandering about in the dark. She worries about falling or getting lost. Darkness is one of the first reasons for Betty to stop driving and she continues to be wary about darkness citing the time she got

lost in a dark room or fell down her friend’s stairs. It is interesting that she seems to blame herself for any problems she has experienced stating she should have thought to put a light on. Harry also recognises that she is liable to get disoriented and more confused where there are low light levels.

<i>“it was dark, and I thought well I’m just coming out of the loo, I’d got up fine I just forgot that there was a little bit of a turn and that was what it was. It was my fault in a way, but I don’t think I knew where the light was.” (Betty)</i>	Identifier: PCA1.33
<i>“I should have had a light on, it was dark, yes, but you see once I’d got out of bed and moved and thought I know that there is a chair there, this is in my mind, and I walked forward and there wasn’t a chair there so then I didn’t know where I was in the system.” (Betty)</i>	Identifier: PCA1.34
<i>“... it’s light and you know where you are going but I think if it is dark you are more likely to get into a panic, definitely.” (Betty)</i>	Identifier: PCA1.35
<i>“It was dark, and she came into the room and she got hold of me you could see relief written all over her face.” (Harry)</i>	Identifier: C1.25

4.2.5.8.4 Colour and Contrast

Neither Betty or Harry think that different colours make a difference to Betty’s visual challenges although when telling me about losing things, Betty does admit that if were put on a similar colour it was difficult to see them (for example: brown rimmed spectacles on a brown table). This seems to be more of a contrast issue than colour problem.

<i>“Like (picks up glasses) these I put them there and then not very long later although sometimes it is a long time as I haven’t found them. ...it’s the colour isn’t it. Dark glasses on a dark table. ... Things disappear.” (Betty)</i>	Identifier: PCA1.36
<i>“I’ve never thought about colour. No, no, it’s never been an issue that suddenly struck me as being co-incidental or significant. Any colour related incidents, I can’t</i>	Identifier: C1.26

<i>remember any? She's always wanted a bright red coat and she got one and was delighted with that. Does she like certain colours, could be, could be red because on the photo you showed her what she saw was the No entry sign where the T bar was red." (Harry)</i>	
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Comment: Both Betty and Harry are aware that Betty has problems seeing properly in low light levels and can become disorientated. It is interesting to note that neither of them has found the use of particular colour helpful for Betty's vision. It appears that good contrast seems more important.

4.2.5.8.5 The use of devices to promote independence

Betty and Harry, with the help of their children, have sourced several products that help Betty, namely an accessible phone, talking watch and the use of talking books. Nevertheless, when asked if he has considered accessible products or gadgets for visually impaired people that might help Betty, Harry appears sceptical:

<i>"... the problem with me, my perception, is that if you get these things, it shortens the time that they have to accommodate these weaknesses. ... Oh, look what's here, it's a cup with two handles, I must be perceived as needing it," (Harry)</i>	Identifier: C1.27
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Betty is asked if she thought the use of an iPad or computer would be beneficial, for instance in helping her to write, but she is reticent to try them, and her diminished level of confidence to learn new things shows in her response.

<i>"... I did read it is one of the hardest things to do when you've got trouble like me to learn some new things because is it worth the torment of it when I can get other people to do it for me. I know it how it sounds, you could think I was a loser, but I just think, I don't think I would be able to learn. (Betty)</i>	Identifier: PCA1.37
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She also speaks of the weariness she sometimes feels trying to do things.

<p><i>“You know that 99% of the time is spent struggling and you think does it matter, does it matter. I do get tired with it all.” (Betty)</i></p>	<p>Identifier: PCA1.38</p>
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Comment: Betty is currently using some gadgets to help her with every day activities but hasn't tried many products specifically designed for visually impaired individuals or people with reduced motor skills. Harry appears reluctant for Betty to use them and infers that using them will make her think she is more disabled than she currently is. He described earlier on in our interview that she has a problem eating but dismisses the use of 'dignity' crockery and cutlery (such as a two handled mug) which might help her eat more independently.

Betty describes vividly how much she struggles to do things and her negative comments 'you could think I was a loser' and 'does it matter' may mean she is starting to give up trying to remain independent.

4.2.6 Support networks and their understanding of PCA

4.2.6.1 Family

Betty and Harry have strong social relationships with both friends and family. Unfortunately, their daughter lives a long distance away, but she tries to meet up with them every few months and keeps in touch with her mum in between visits via the telephone. The couple are looking forward to the birth of their first grandchild. Their two boys live nearer and visit more often. The couple stress their wish to carry on as normally as possible around the family and try to hide any difficulties from them.

<p><i>“The family? Oh, they are wonderful. We just carry on. They say, ‘are you alright?’ and I just say ‘yes’ (laughs)” (Betty)</i></p>	<p>Identifier: PCA1.39</p>
<p><i>“...the boys say, ‘oh I don’t know how you do it, you’re marvellous’. I’m not...There’s nothing marvellous about it.” (Harry)</i></p>	<p>Identifier: C1.28</p>

The family are very aware of the difficulties that the couple are experiencing and try to help in practical ways. The boys are sorting out easy dial telephone numbers on an accessible phone for Betty and her

daughter provided a talking watch when she found out her mum was having difficulty in telling the time.

4.2.6.2 Friends

The couple have a small group of friends who they have known for many years and they meet up regularly. All the members of the group are living with various long-term conditions but support each other.

<p><i>“...we’re disintegrating. The lawyer, his wife died of cancer, dreadful, he has a terrible back, his spine. It’s an inoperable condition. Another friend has motor neurone. The last couple, well he had a heart attack out of the blue. His wife has been on steroids for years for some internal condition. She’s the youngest of us all. We used to say when we were young by gum you’ve got a lot of funerals to go to in the days when it was a joke. It’s not a joke now. But as you become older you then become the behaviour of older people caring together.” (Harry)</i></p>	<p>Identifier: C1.29</p>
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Outside of their close friendship network, Harry is rather scathing of the ‘advice’ given by other people who do not fully understand their circumstances:

<p><i>“... particularly with PCA you get the type ‘well she looks alright. Yes, she must be alright surely’. And you get all kinds of tips from people.” (Harry)</i></p> <p><i>“Let me say this, it’s rather rude I know but in that department there are no shortage of anal ventriloquists (laughs). They just don’t know, and you have to be so forbearing and so patient because some of these people are friends. They mean well. (laughs). How many silly things are done under the banner of ‘meant well?’” (Harry)</i></p>	<p>Identifier: C1.30</p>
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4.2.6.3 Health and Social Care Professionals

Apart from a cleaner who comes in twice a week, Betty and Harry have no other professional assistance. Harry describes a social worker that came once to offer support. He does not think it was of any use as the person seemed unaware of the type of difficulties that Betty was experiencing:

<i>“...older people’s office or something, they called, and they chatted. Very nice lady, very helpful, but it became obvious that what she was looking for was Zimmer frames and handles in the bath and this sort of thing more than anything else and I didn’t press the point” (Harry).</i>	Identifier: C1.31
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Harry has had a carers assessment and they had advised him to have a break from caring for his own personal health, but he is finding it difficult to do this:

<i>“the carer, my carer lady, lovely girl said ‘never mind about the patient, let’s think about you. You’ve got to get out on your own. You’ve got to laugh, argue, drink, whatever. You must get away’. (pauses), You can’t! She says I’m spending too much time with Betty. (Harry)</i>	Identifier: C1.32
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<i>“I think there is a sense of isolation due to the rareness of the condition” (Harry)</i>	Identifier: C1.33
<i>“If you have recourse to the GP in the majority of cases, [they], don’t understand PCA. If you go to a counsellor, they most certainly don’t understand PCA.” (Harry)</i>	Identifier: C1.34

When asked about day centres, he is dismissive:

<i>“Yes, I have thought about day centres but there will be people there, you know, somebody playing the piano and somebody playing cards and it can be a bit counterproductive. This is not being snobby that’s just being realistic because PCA is different. (Harry)</i>	Identifier: C1.35
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However, he expresses enthusiasm about the thought of a support group for other people with PCA that is near enough to attend.

<i>“It would be nice to meet up with (name) whose husband has PCA and (name) from (city) and (name) from (city) because their wives have PCA and we know what each other are going through.” (Harry)</i>	Identifier: C1.36
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<p><i>“...I think it would. I hope it would... I think that people that are able to cope with it and people who aren’t able to cope with it, if they interface, some of the confidence might rub off on them and they will know. ... The loneliness and isolation is reduced by these meetings. You are not alone.” (Harry)</i></p>	<p>Identifier: C1.37</p>
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Comment: Betty and Harry continue to have good social contacts with friends and family. However, some friends don’t really understand their circumstances and can give inappropriate advice which annoys Harry. The couple have also found health and social care professionals lack awareness and understanding about PCA and how to support Betty’s specific needs. Harry appears to feel unsupported and lacks people to talk with who understand his situation and he longs for a PCA Support Group that is near enough to attend so he could meet likeminded individuals.

4.2.7 Changing roles within the couple’s relationship

The difficulties faced by Betty due to PCA has impacted and changed the dynamics within the household. Betty admits that:

<p><i>“Harry’s doing lots of things he would never have done. I mean I took it that as women’s work and I think most women of certain ages, you look after your family and the husband looks after the rest. Harry had never ironed a shirt. He didn’t change any nappies but that didn’t bother me, so I suppose there are things that he’s had to do that he wouldn’t have expected to do.” (Betty)</i></p>	<p>Identifier: PCA1.40</p>
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Harry expands on how his life had changed:

<p><i>“... the word challenge does sum it up. Your responsibilities have become tremendously wider ranged. I do all the shopping. I do all the cooking. I pay the bills. I’ve dualized all the bank accounts and I’ve got two powers of attorney, property and health and welfare. ... You can imagine the everyday things that she finds impossible to do, I just do it.” (Harry)</i></p>	<p>Identifier: C1.38</p>
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When asked how he feels about the change in his role, Harry is matter-of-fact about the situation:

<i>“It’s really irrespective on how I feel on a particular day. It’s there to be done. That’s how I handle it for me. It’s do it, do it properly, do it the same way, make a habit of doing it every time. ... you can just become rather boringly well-disciplined, but it makes life easier for me to the point where if Betty comes into the kitchen, I know she’s been in because tops are left off, drawers are open which just a little bit, I know It’s sad, but just a bit irritating.” (Harry)</i>	Identifier: C1.39
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4.2.8 Wanting to protect the person with PCA

Harry has also become very protective of Betty, trying to shield her from harm and distress.

<i>“I think PCA patients need protecting to a degree. There’s only a certain amount that they can do on their own. What they do on their own is very credible, very remarkable. But they do need protecting from their own vulnerabilities and weaknesses or the effects of their own vulnerabilities and weaknesses.” (Harry)</i>	Identifier: C1.40
<i>“... it’s best when the first suggestion that there are difficulties there to be stopped stone dead because it is not going to get better, it’s going to get worse.” (Harry)</i>	Identifier: C1.41

Unfortunately, in the process, he is taking things that Betty loves doing away from her which then causes her frustration and upset.

<i>“I bought her a colouring book, she couldn’t do it, she was going way over the lines. I noticed three of them. So, in order that she wouldn’t be frustrated or depressed I took the book and hid it and it is still hidden. I took it off her.” (Harry)</i>	Identifier: C1.42
<i>“I wouldn’t let her out on her own now. I wouldn’t even let her go to the town on her own.” (Harry)</i>	Identifier: C1.43
<i>“She doesn’t do any cooking or baking. None whatsoever.” (Harry)</i>	Identifier: C1.44

But discussion with Betty shows the lack of stimulation is starting to dent Betty’s confidence and esteem further:

<i>“I feel I am wasting time, wasting time. I wish I had something which would be useful but every time I touch anything it comes to grief it seems.” (Betty)</i>	Identifier: PCA1.41
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4.2.9 The emotional impact of living with PCA

Betty and Harry have been together for over 50 years and their love for each other is clear to see. Harry appears unemotional and in control throughout the interview until I ask how he is coping with the situation. At this point, he becomes very tearful but says he wishes to continue talking with me. He explains the pain of watching Betty change in personality and feeling helpless to stop it:

<p><i>“There is the perception with PCA carers that they are watching someone who they love, who they have been married to for thousands of years. That they’ve walked hand in hand on beaches with deliberately avoiding the obvious almost. That’s what it appears. It appears in some way that you are living with a doppelganger. Somebody who is not the person that you married. Somebody in a different form. I don’t want the different form. It’s almost like a (pause) like an image not (pause) Doppelganger, complete doppelganger. Who are you? Where’s the real you? Where’s the girl I could have a normal conversation with? ... It’s gone, gone, long gone. (Harry)</i></p>	<p>Identifier: C1.45</p>
<p><i>“I feel very sad, very, very, very sad. Mixture of anger at the situation, anger with myself because I can’t handle it. Great sadness. In fact, a dreadful sadness. There is no absence of tears.” (Harry)</i></p>	<p>Identifier: C1.46</p>
<p><i>“.. the black dog I get that, yes. But I’ve always had it you see. It’s nothing new. But it can be caused by relatively minor things. It must be made from a cocktail of events, cocktail of situations.” (Harry)</i></p>	<p>Identifier: C1.47</p>

When asked the same question, Betty appears resigned to her condition but also shows moments of frustration and anger when she talks about how she is feeling:

<p><i>“...I’m aware that there is no cure, so I’ve got to accept how it is and that’s it. ... I wish it hadn’t happened obviously. I mean it has turned our lives upside down. (Betty)</i></p>	<p>Identifier: PCA1.42</p>
<p><i>“...I know people have to be ill but why did have to be me. ... at least I haven’t got pain” (Betty)</i></p>	<p>Identifier: PCA1.43</p>
<p><i>“... I’m just frustrated. I feel as if something is eating me up. There’s more bits coming off as it were.” (Betty)</i></p>	<p>Identifier: PCA1.44</p>
<p><i>“I suppose you could say I feel cheated”. (Betty)</i></p>	<p>Identifier: PCA1.45</p>
<p><i>“I do get sad, but I think I could be sadder. I’m not looking to be sadder.” (Betty)</i></p>	<p>Identifier: PCA1.46</p>

Comment: Harry and Betty both express sadness about their situation. Harry speaks so candidly about feeling isolated and having episodes of 'the black dog'. Some 'time out' from his caring role may benefit his own health. Betty appears unhappy but perhaps her antidepressant medication is masking her true feelings. One moment she speaks resignedly about her situation, the next she expresses frustration and some anger. Her expressions 'something is eating me up' and 'bits coming off' seem to imply she feels she is disappearing as a person.

4.2.10 Facing the future

When asked about the future, Betty shows a concern about losing her identity, describing how she looks in the mirror to check she still can recognise herself. She explains:

"I think you tend to think of people who are ill, because I am technically ill, that it shows in your face and that you're going to be a wreck. But I look, and I think, yes it's me." (Betty)

Identifier:
PCA1.47

When asked what she is fearful of, she clarifies:

"Well I hope I won't get too much changed to how I am now. I don't want to become a stranger to myself, I suppose" (Betty)

Identifier:
PCA1.48

Harry, on-the-other-hand, describes his thoughts of the future more pragmatically:

"I think about the future from a financial aspect. At what point will I need to put Betty into care? Need, not want to, because it will be heart breaking. We are going to need something like 60 to 70,000 pounds a year and so say the time within a care home is two years, that's £100,000 plus that you've got to hide in a sock somewhere." (Harry)

Identifier:
C1.48

Although he finishes our conversation wistfully

<p><i>“I don’t want to get to the situation when I’m helping her along corridors, feeding her with a spoon and this and that. It will be humiliating for her.” (Harry)</i></p>	<p>Identifier: C1.49</p>
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Comment: *Betty and Harry appear to view the future differently. Betty focuses on her condition. She does not mention long term care possibilities at all. On the other hand, this is clearly on Harry’s mind and he appears resigned to one day having to put Betty into a Care Home and the financial costs of doing this.*

4.2.11 Summary of Betty and Harry’s Story

Betty presents with several PCA symptoms as shown in [Table 16](#) below .

These include:

Table 16: Betty's presenting PCA symptoms

●	Acalculia
●	Agraphia
●	Alexia
●	Apraxia
●	Associative agnosia
●	Contrast sensitivity
●	Depth perception difficulties
●	Dressing apraxia
●	Environmental agnosia
●	Heightened sensitivity to glare
●	Left sided visual neglect
●	Ocular motor apraxia
●	Simultanagnosia
●	Visual disorientation

The couple describe how minor incidences occurring to Betty over many years which they put down to the process of getting older. Betty first noticed she had a problem with driving because she could not work out how close she was to adjacent cars and following bumping the car in the garage she visited the opticians who did not appear worried about her sight. This resulted in Betty continuing to drive for several years even though her vision was getting worse.

When the GP finally referred Betty to hospital for further tests, her diagnosis was delayed because she did not fit into the normal bracket of having 'dementia'. Her memory was good and at first her brain scans did not show any atrophy. Now diagnosed, the couple appear to take this result pragmatically and are working together as a team to manage the condition.

A serious fall down a friend's stairway has resulted in Betty being very afraid of falling. This has been compounded by further falls when out walking or sitting down due to left side visual neglect and depth perception problems. Harry does not seem to realise how afraid Betty is of falling and thinks her friends compensate for her too much by helping her sit safely as well as not considering the need for hand rails in the bathroom. When descending stairs, Betty describes a strange perception of something 'pulling me down' which causes her to panic that she will fall. She uses words like 'frightening', 'panicky' and 'torture' to describe her feelings.

Betty now finds it impossible to read, write her name, identify products such as shampoo, food stuffs, and understand money. Dressing is very difficult, and she usually needs the support of Harry to do this successfully.

Although she loves to cook and bake, Harry doesn't let her do this anymore because of her inability to read labels and recipes and her poor motor skills in trying to use baking and cooking equipment. By doing this he feels he is protecting her from harm or being distressed if she was unable to undertake a task successfully. However, Betty revealed feelings of uselessness and boredom. They have not tried any of the RNIB products that help blind and visually disabled people to undertake these tasks.

The couple still socialise with old friends although Betty finds it hard to use cutlery and has left sided visual neglect so often misses food on the left side of her plate. Harry realises that Betty has a problem eating but dismisses the use of adapted crockery (such as a two handled mug) which might help her manage more independently.

Betty has got lost both outside and in her own home particularly if there are low light levels. When walking certain surfaces are problematic such as

shiny or patterned surfaces because of her heightened sensitivity to glare and contrast and depth perception problems.

Betty's energy levels are low. She explains it takes a great deal of effort to do things and this tires her both mentally and physically. Even though she has been having vision and perceptual difficulties for 14 years, her memory remains quite good although they have both noticed her short-term memory has recently started to deteriorate.

Betty describes vividly how much she struggles to do things and her negative comments '*you could think I was a loser*' and '*does it matter*' may mean she is starting to give up trying to remain independent.

Although, Betty and Harry continue to have good social contacts with friends and family. Harry thinks that some friends don't really understand their circumstances and can give inappropriate advice which annoys him. They have found health and social care professionals lack awareness and understanding about PCA and how to support Betty's specific needs. Harry appears to feel unsupported and lacks people to talk with who understand his situation and he longs for a PCA Support Group that is near enough to attend so he could meet likeminded individuals.

The couple's individual roles have changed within the household. Harry appears to manage quite well but has taken full control of running the home and it is clear he wants to keep Betty from harming herself. He has also taken away her role in life, her identity of being a wife and homemaker so when she tries to help or messes up his routine he admits to being frustrated with her so if she can't do something independently he takes it away from her (e.g. the colouring book, going out, cooking and baking). Although this seems to be unintentional, Harry is undermining her personhood and decreasing her state of wellbeing although he does it under the banner of 'protecting' her and enabling him to cope with his caring role.

Harry and Betty are both showing symptoms of unhappiness. Harry is resisting social care for Betty (day centre) at this point. However, because he spoke so candidly about feeling isolated and having episodes of '*the*

black dog, he does require some *'time out'* from his caring role for his own health. Betty is quite sad, but her antidepressant medication may be masking her true feelings. If some intervention could be found to enable her to do some of the hobbies she loves (e.g. baking, art, shopping) her general wellbeing may improve. A day centre or respite facility that is fully aware of her specific visual and perceptual difficulties may help the couple in different ways.

Betty and Harry remain a strong loving couple, though they view the future differently. Betty is thinking about her condition and worries if PCA will change her so much she will not recognise herself. She does not mention long term care possibilities at all. On the other hand, this is clearly on Harry's mind and he appears resigned to one day having to put Betty into a Care Home, the costs of doing this and the emotional upheaval this will cause to them both.

4.3 Case Study Two: Polly and Joe's story

Polly and Joe were interviewed at home, alone in separate rooms to enable complete privacy. Each interview took one hour, was tape recorded and fully transcribed. Polly expressed great concern that her interview would not be totally confidential, and I reassured her that Joe will not read the transcript of our conversation and that all names and places will be anonymised.

4.3.1 Setting the Scene

Polly is a 73-year-old lady who had been diagnosed with PCA for 6 years. She appears a bubbly and forthright person. Polly has been married to *Joe* for 43 years. It is her second marriage. Her first husband had died when he was a young man leaving Polly with two baby daughters to bring up alone. One of the girls died due to cot death. She and Joe live in *Appleton*. They moved elsewhere for a number of years. Polly worked as a welfare officer supporting mothers and children. She had been caught up in a bombing incident whilst out shopping but had not sustain serious injury. During this time, her general health was good although she had suffered from poor sight from childhood. Her health had deteriorated as she grew older. Apart from PCA, Polly now suffers from high blood pressure, glaucoma, diabetes, emphysema and psoriasis which she controls with medication. She has had a total hip replacement and cataract operation on her right eye. Joe is 69 years old. He left school at 15 years of age and was employed as a factory worker. When the couple moved , Joe elsewhere worked as a factory manager. During this time the couple described their lifestyle as fun and exciting. They had servants and enjoyed entertaining with barbeques and pool parties most weekends. Unfortunately, when they returned to England, Joe was unable to get paid employment, so he took on voluntary roles within the local community. Joe advises that he did not suffer from any long-term conditions although he does admit to being short tempered and constantly stressed.

The couple now live in a rented house near their daughter. It has good natural lighting throughout. There is a small lounge containing a sofa, armchair and television. The kitchen is small with a cooker, microwave and fridge/freezer. It also has a small kitchen table with two chairs. Polly has a bedroom downstairs next to an accessible bathroom with shower and toilet. Joe has a bedroom upstairs. Carpets and the décor are plain. The walls are covered in photographs of the couple’s married life. There is also a small garden consisting of a lawn and flower beds.

4.3.2 Realising something was wrong

Both Polly and Joe had felt that something was going wrong with Polly’s vision more than forty years previously.

<p><i>“We thought her sight wasn’t fantastic, but she could still see. I would say back in the 70s cause I was with her when she used to go to the doctors and the doctors would just pass her off. ... but it’s one of those things that ... rubbing her eyes one night she couldn’t see and then it come back and then it happened to again and we went to this guy and he said ‘look you might lose your sight and we don’t know when it’s going to happen and it might come back in an hour or next week or three months or never’. He said, ‘I don’t know, and we don’t know when it’s going to happen’. ... that was back in the 70s.” (Joe)</i></p>	<p><i>Identifier: C2.1</i></p>
<p><i>“I can’t remember. ... my eyesight has been bad for years. From being a young girl, I’ve always had bad eyes and then when this thing started.” (Polly)</i></p>	<p><i>Identifier: PCA2.1</i></p>

Polly was given some tablets and cortisol injections. They weren’t sure what they were for, but her eyesight seemed to improve a little and she could still see. In 1986, the couple moved elsewhere but after about three or four years, Polly had another isolated incident of vision loss, but seemed to recover and she continued driving her car.

<p><i>“She was driving her car although she only used to go in the close proximity. She didn’t venture far, and I always knew she had poor sight, but she was still able to drive a car.” (Joe)</i></p>	<p><i>Identifier: C2.2</i></p>
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However, she was concerned and so went to see an optician.

<i>“me vision was just getting worse and again, you see, I kept going to the opticians and they kept saying there was nothing wrong. When we was in [place], no, it wasn’t noted or anything.” (Polly)</i>	Identifier: PCA2.2
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Polly and Joe saw another doctor who encouraged them to seek advice when they got back to the UK.

<i>“The guy He would say to Polly; do you know I am pleased you are going back to England because there’s summat wrong. (Joe)</i>	Identifier: C2.3
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Polly described why she stopped driving once they went back to England:

<i>“I just couldn’t see. I couldn’t gauge it properly. I stopped myself from driving. I was a danger on the road. You know when we got off the plane ... I said to Joe ‘that’s it. I daren’t go on the traffic here because things would just come at you. You don’t know they’re there’. I just give it all up as soon as I got here. I missed it. I missed it a lot. It was a lot of my freedom.” (Polly)</i>	Identifier: PCA2.3
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Comment: Polly and Joe describe her sight as being poor for many years and they appeared not to know the cause for this at this point. It is interesting that she feels her driving is dangerous. When she describes that cars ‘come at you’ and ‘you don’t know they’re there’, is this because she is losing peripheral vision? Such problems may be due to PCA, but they could also be the result of glaucoma?

Polly also recognises that stopping driving has diminish her independence. She talks about losing ‘my freedom’.

4.3.3 Journey to diagnosis

In 2007, when they returned to England, Polly was referred to an eye clinic. She was initially told that she was suffering from glaucoma and that was

the reason for her eye problems, only to be told a few months later that she didn't have the condition. The couple were perplexed.

<i>“Well to be quite honest it can be quite confusing... not just for me but for Polly as well cause we'd come out and say 'do you know what he bloody said? He said I've got it and you go a few months later, you haven't got it and then you have got it and I think it's because they don't understand what PCA is” (Joe)</i>	Identifier: C2.4
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During one of these checks an optician thought there was something wrong and referred her to the consultant specialist at the eye hospital who then referred her on to a neurologist.

<i>“It was one of the opticians. We saw him at the eye clinic at the hospital and he turned round and said 'just a minute there's something wrong here ... and then we was referred to the top guy at the eye clinic ... but he couldn't explain why when she'd go for field tests or whatever so he referred us to a neurologist who put her through these memory tests.” (Joe)</i>	Identifier: C2.5
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Polly was finally diagnosed with PCA in 2011.

Comment: *There was a delay of four years from being referred to the eye hospital in the UK and getting a diagnosis of PCA. The cause of her sight problems appeared to be glaucoma in the first instance, so it took time to finally refer her on to a neurologist.*

4.3.4 Reactions to knowing

Polly describes the fear she felt when she heard the diagnosis of PCA and how she thought the specialist was not sympathetic:

<i>“The first thing I thought was ‘Oh I'm going blind. That was your first thing, the fear of going blind. Looking at my daughter and thinking I'll not see her. (Polly)</i>	Identifier: PCA2.4
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A qualitative study exploring the specific everyday social and environmental challenges faced by community dwelling people living with posterior cortical atrophy (PCA)

<i>“He didn’t explain. He was awful that guy. I said to him one day I still didn’t know what it was. I said to him, ‘I’m going blind’. ‘No, you’re not, you’re not’. I said, ‘I know I am’.” (Polly)</i>	Identifier: PCA2.5
<i>“I was upset because right from the beginning they kept saying ‘there’s nothing we can do. There’s nothing we can do’. Everybody says the same.” (Polly)</i>	Identifier: PCA2.6

After reflection, Polly say she started to feel more positive:

<i>“... when they told me that it wasn’t a brain tumour then I had a bit of relief. (Polly)</i>	Identifier: PCA2.7
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Joe, on one hand, says he found it helpful to know what the condition was, but at the same time was appalled with the way one of the specialist had spoken to them during a consultation.

<i>“The brain. I’d never thought about that previously” (Joe)</i>	Identifier: C2.6
<i>“That psychiatrist. ... my wife was sat there, I was sat here, he looked at me and he said, ‘do you know Joe, you’ll have to learn to lie’. I said, ‘I beg your pardon?’. He said ‘Like politicians, you know if you ask them a question they never answer it. ... Well, if your wife asks you something you have to learn to lie. Answer her but don’t answer her, lie.’ And Polly is sat there, and she heard this. ... and I’m thinking for God’s sake she’s not deaf! ... and, of course, when we walked out, she said ‘I don’t like that man’. Well I can understand. And how the hell can I explain and if I ever said aught. ‘are you telling me the truth?’ so it’s in her brain cause he told me and she’s thinking it.” (Joe)</i>	Identifier: C2.7

Comment: The way the couple describe being given the diagnosis of PCA is a cause for concern. From Polly’s perspective she was not given any emotional support from the health care team. Joe’s anger at the way the psychiatrist spoke to him in front of Polly is understandable. The psychiatrist dismisses Polly as a person in her own right, talking to Joe in front of her as if she doesn’t understand what he was saying. This goes against all person-centred approaches to supporting people with dementia. It is worrying to know that this type of behaviour still exists within healthcare staff in the UK.

4.3.5 Day to Day challenges

Polly struggles to undertake many activities of daily living and needs Joe to help her with most tasks.

4.3.5.1 Personal Care

Polly cannot recognise washing products so, apart from a regular trip to the hairdressers, Joe needs to assist her in washing and personal care.

<i>“She cannot recognise washing stuff. I have to place them there. We’ve got one we use which is like a soap substitute, but it looks a bit oily, so I say your soap’s there, your sponge is there. She has a standing appointment with the hairdresser, so they wash her hair. She used to have a hairdressing and clothes business although she’s not a qualified hair dresser, but she’s always done her own hair and that was another thing, curling and doing all that, we had to change that cause I’m not very good at that” (Joe)</i>	Identifier: C2.8
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Comment: *Not recognising the washing products could be because Polly has lost the skill of reading words (Alexia) and cannot read the labels. Because Joe describes her as not recognising washing stuff, this could be a symptom of [associative agnosia](#) if she is unable to identify simple objects even though she knows what soap and shampoo are.*

Polly does not like Joe helping her with personal care. From our conversation, it appears she dislikes him touching her in intimate areas.

<i>“I stand in the shower with my back to Joe and then he’ll wash all me back. You have to go gentle ...I’ve got terrible skin. I know he’s my husband, but I don’t like (pause) you know when you’re doing the bit between your legs. I don’t like him doing that even though he’s me husband. There’s little things I don’t like him doing.” (Polly)</i>	Identifier: PCA2.8
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Nevertheless, she feels it is important to her that she always looks presentable.

<p><i>“I still try and keep myself to look good. I shove a bit of lippy on and then I feel done up myself, not scruffy.” (Polly)</i></p>	<p>Identifier: PCA2.9</p>
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Comment: Polly demonstrates signs of distress over Joe touching her in intimate places. Her personal identity shines through when she describes wanting to still look good.

4.3.5.2 Dressing

Polly admits she finds getting dressed difficult but states she can manage if she takes her time although putting on a bra is impossible.

<p><i>“I used to be able to stand up and pull a sock on. Now I have to sit down ... but otherwise I really think I can do most of it when I’m taking care is because of what I’ve got that I will slow myself down to do it.” (Polly)</i></p>	<p>Identifier: PCA2.10</p>
<p><i>“I can’t do bra. ... I do my trousers and everything like that, it’s just the brassieres.” (Polly)</i></p>	<p>Identifier: PCA2.11</p>

Joe, on the other hand, states that Polly couldn’t dress herself and he must support her. Interestingly, he mentions that she can probably do it herself, but it would take a long time, then dismisses this by saying that even then she will probably do it wrong.

<p><i>She can’t dress by herself. ... like a top I have to make sure I put it over her head then right arm through and left arm through because sometimes she pushes against it. And then trousers that way round cause often she will put trousers on back to front. ... She might be able to manage herself, but it would take a long time and then the clothes are possibly inside out and back to front. You know you can’t really complain to her because she’s using as much sight as she can.” (Joe)</i></p>	<p>Identifier: C2.9</p>
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Comments: Joe is describing [dressing apraxia](#). Polly believes she can dress herself slowly. Has she totally lost the ability to dress or is it that Joe is not patient enough with her to let her dress at her own pace with help from him when required?

4.3.5.3 Food Preparation

Polly was a great entertainer when she was younger and thoroughly enjoyed cooking and baking. Now she struggles to prepare anything. Reading recipes and packets were impossible but Polly still wants to carry on with the hobby she loves.

<i>“...I can’t read a menu, not a menu. I wouldn’t be able to read it unless someone read it to me and then I would start mixing, doing all those things, you know. Now I would just have a go. I can’t read packets and things, no. Everything has to be fresh” (Polly)</i>	Identifier: PCA2.12
<i>“I would be frightened to put the stove on or put anything into a stove and bring it out. I would be very cautious over that in case I did drop it or summat. I think I would still attempt it if Joe was in the house and he could come and help me (Polly)</i>	Identifier: PCA2.13

However, Polly speaks angrily about Joe, accusing him of not wanting to help her.

<i>“He wouldn’t, there is no teamwork” (Polly)</i>	Identifier: PCA2.14
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Joe explains that the kitchen is too small for two people to be cooking at the same time:

<i>“... she’ll come in and say can I help you and I say to be quite honest love it’s not a large kitchen so sit down.” (Joe)</i>	Identifier: C2.10
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However, after closer questioning, it seems that Joe finds the whole situation very stressful. He resents people telling him he should let Polly prepare food even though he has bought accessible gadgets to try to assist her. He says he is afraid Polly will harm herself or react badly if she does hurt herself; but further on in the conversation it becomes clear that he also resents the mess she makes when she does try to help.

<p><i>“if I say peel potatoes I don’t want to have to start bandaging cuts up because I know it is a (pause) if you cut your finger with Polly she loses it and I try to avoid it or protect her.” (Joe)</i></p>	<p>Identifier: C2.11</p>
<p><i>“I mean last week she said can I help you but A lot of people will say, ‘Oh you should do this, let her do that, let her do this’. She’s got blooming things in there. She’s got a talking scale and a talking measure and we’ve got that yellow thing on the fridge for when you pour water into a cup but why should I turn round and say do it because there would be water slapped, I’ve seen it, water slapped all over the place and I would rather do it myself because I know she is not going to harm herself or burn herself whatever but see it’s easy to maintain as well and I’m the one who has to clear the mess up. .. I say, ‘no I don’t want you harming yourself’. but I think she gets upset because she can’t do this, and she doesn’t do that, but I can understand from her side. ... we’ve just had taps changed and like she went for a drink yesterday and she couldn’t even remember how to operate them. It’s not her fault, I can understand that, but when these things happen you tend to think it’s something like a child might say ‘how do you do that. You have to look at it in a totally different light.” (Joe)</i></p>	<p>Identifier: C2.12</p>

For Polly, not letting her continue her role as ‘housewife’ is deeply upsetting.

<p><i>I hate it (pause), there’s nothing I can do. ... I sit there and have a little cry some days on my own. I’m not ashamed of it. I do it and (pause) maybe I do feel a bit better after.” (Polly)</i></p>	<p>Identifier: PCA2.15</p>
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Comment: Polly’s lack of ability to read (*Alexia*) and reduced motor skills (*apraxia*) in addition to her diminished visual capabilities are causing difficulties with cooking and baking. Joe says they have many RNIB talking products that could help her in this situation but is reluctant to let her use them because she will make a mess or hurt herself. Polly describes their lack of ‘team work’ and to enable her to cook and bake they would need to work together. Joe is finding this situation stressful because it creates more work for him. Hearing Polly talk about crying because she is missing her housewife role demonstrates her emotional wellbeing is being compromised.

4.3.5.4 Eating and drinking

Joe describes the challenges Polly has in using cutlery but states that she tries her best to remain independent.

<i>“Many times, I have to cut the food up but at other times she’s quite independent and she’ll say, ‘no I can do it myself’. She’s got one of them blue bibs that she puts on to protect tops and everything. Just puts it on at meal times.” (Joe)</i>	Identifier: C2.13
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Joe needs to ensure Polly takes her medication. This needs to be supervised, (although Polly does not like this), because she finds difficulty in seeing them.

<i>“Like when there are pills there I pick her hand up and put them on the pills. If the pills were there she wouldn’t see them, so I say give us your hand and I put it there. Initially she can be quite resistant because I am taking her to them but it’s the only way or she’d be all over the place” (Joe)</i>	Identifier: C2.16
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Comment: Polly’s diminishing motor skills are causing her problems with her ability to eat independently. Joe talks about having to give medication to Polly because she ‘wouldn’t see them’. Is this because she is not able to see a whole scene at once and is only perceiving individual elements and so misses where Joe places them (*simultanagnosia*)? Her peripheral vision may be compromised due to glaucoma or visual neglect caused by PCA. Would putting them in a brightly contrasting container directly in front of her help to find them?

4.3.5.5 Reading and numeracy

Polly’s reading and writing abilities have deteriorated to the stage where she can no longer do these tasks. Still trying her best to be independent, Polly describes how she will try anything she can to improve her eyesight but up to now nothing has helped:

<p><i>“... I can’t sit and write anything. I can’t read anything as what I could in them days, in them early days. I just used to look more, and I used to go and get special glasses and that. I’ll go anywhere if they say there is something you can try. I’d go and try everything there is.” (Polly)</i></p>	<p>Identifier: PCA2.17</p>
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The couple have tried talking books but found the narration not to their taste:

<p><i>“I did try talking books, but we found them so pathetic, old fashioned ways of everything. We didn’t like that. Joe tried as well.” (Polly)</i></p>	<p>Identifier: PCA2.18</p>
<p><i>“She used to enjoy reading and we’ve even tried these talking newspapers and books. Oh crumbs. You get this woman (puts on a posh voice). Hello, this is about so and so in the park. And then there will be a romance and it’s the same bloody woman.” (Joe)</i></p>	<p>Identifier: C2.15</p>

Polly always used to manage the housekeeping finances, but she now is unable to count or recognise money. She reminisces about being a housewife in her earlier life and is resentful that Joe now does this task.

<p><i>“Every housewife does this I’m sure no matter what’s gone on for the month, at month end you cover your shopping and I used to love that day because there was such beautiful food to buy. And he’s took that off me and I tried to do it nicely to him” (Polly)</i></p>	<p>Identifier: PCA2.19</p>
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Joe seems unaware of Polly’s feelings of loss and states that he still gives her money every month so that she is able to treat herself.

<p><i>“...I gave her £160 the other day cause it was the end of the month so I do it that way so she has got a little bit of independence but she can’t tell the difference between £5, £10 or £20.” (Joe)</i></p>	<p>Identifier: C2.16</p>
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Comment: Polly and Joe are describing symptoms of *acalculia*, *agraphia*, *alexia*. Polly's expression 'I'd go and try everything', reveals her strong desire to find a solution as well as her feelings of helplessness over the situation. Her loss of identity as a housewife comes across strongly when she talks about the Joe taking over her role. However, she is blaming Joe for this situation rather than considering her decreasing visual and perceptual abilities.

4.3.5.6 Problems with Perception

I ask Polly to describe her vision. She understands that she didn't see things exactly as they are and explains how this affects her.

<i>"It's not blurry. I can still pick out things, but you look at something and you don't see it. (Pauses). You do see it, but you're not seeing a true picture and you know you are not seeing a true picture of things and you wish you could wipe that away. But I can look at my daughter and I can see (pause) well still think I do think I do see her right but obviously I'm not but I'd give the world to see her" (Polly)</i>	Identifier: PCA2.20
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Now that she couldn't read anymore, Polly states she gets a lot of pleasure from watching TV.

<i>"I watch TV, yes. I can follow it, yes" (Polly)</i>	Identifier: PCA2.21
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But Joe has a different opinion. He has obtained spectacles from RNIB to try and help her watch TV but feels that her concentration, rather than her sight, is the problem.

<i>"I don't think she is seeing the TV properly ... and it's a decent size telly. I've got her different coloured glasses that you can get from the Blind Institute depending on your eye condition, it's a different colour. ... but a lot of the time I feel that it's her attention span." (Joe)</i>	Identifier: C2.17
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Comment: Polly is aware she is not perceiving people, objects, scenes correctly (*Simultanagnosia*). Television appears to be enjoyable for her although Joe is less sure she can see the picture properly.

Joe also expresses frustration and despair when sometimes Polly doesn't recognise him.

<i>"She's turned round and said 'Who are you?' Like often, ..., 'who are you?' or she would say 'oh Joe wouldn't have done that', I say 'hey who the hell am I?'. And you know ... when somebody says that to you, you tend to think, is there any hope. (Joe)</i>	Identifier: C2.18
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He also describes frequent incidents when she has not recognised their home.

<i>"...and then she'll say how long have you had that house. I say it not my house, it's our house and it's a rented house. No its not a rented house. ... I want to go back to my other house. We had all that a few months ago" (Joe)</i>	Identifier: C2.19
<i>"We are home. No this isn't my home. I said its (tells address). She says it's not. I showed her the 15 on the door. It happens, I reckon about two or three times, ... a week. It can last about 3 or 4 hours." (Joe)</i>	Identifier: C2.20

Comment: Joe is describing *prosopagnosia* (face blindness) and *environmental agnosia*. These symptoms seem to be currently transient as Polly is not experiencing them all the time. However, they appear to be quite frequent and Joe describes feelings of distress and helplessness when they occur.

4.3.5.7 Memory and insight

Polly thinks her memory is OK in general but during our conversation advises that she finds it difficult to respond sometimes to my questioning.

<i>“Everyday memory. Just me doing the house and going around and that I don’t think it’s bad at all. It’s cause you’re asking questions when I’ve got to think of an answer now it’s not same.” (Polly)</i>	Identifier: PCA2.22
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But Joe thinks that Polly often forgets where she puts things, often accusing people of stealing.

<i>“She forgets where she puts things so often she’ll say somebody has pinched me money and I’ve had to turn stuff upside down.” (Joe)</i>	Identifier: C2.21
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<i>Comment: Our conversation makes Polly feel that her memory is not as good as she first thought. Joe describes her as forgetting where she has put things. This could be forgetfulness and/or the fact that she cannot find things due to her vision and perception difficulties.</i>
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4.3.5.8 Environmental challenges

The physical environment also causes Polly difficulties either with physically moving around or correctly perceiving what is in front of her.

4.3.5.8.1 Balance, walking and fear of falling

Polly admits that her energy levels are low which Joe puts down to the fact that she has never been an athletic person.

<i>“I’m not the same as I used to be, not as much energy” (Polly)</i>	Identifier: PCA2.23
<i>“... it’s not as though she can walk far, she says oh, her legs are hurting this that and the other.” (Joe)</i>	Identifier: C2.22

Following her hip operation, Joe feels Polly's balance has become poor.

<p><i>“To be quite honest, her balance is poor since she’s had her hip operation. She sways. I just say, you were born on the side of a bloody mountain you’re cockeyed one way but it’s after having that hip replacement. I have also noticed now she shuffles more than taking steps” (Joe)</i></p>	<p>Identifier: C2.23</p>
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These problems, in addition to her vision difficulties due to PCA, have caused Polly to fall when out walking on pavements. They have tried a walking stick which hadn't help so Joe has resorted to holding on to her when they are out.

<p><i>“She has a walking stick but because her sight’s not too good and the pavements are not too good as well. When we’ve gone out, she’s actually misplaced it and she fell and I said, ‘be buggered to that and just hang on to me arm’ and I’ll say, ‘step up, step down’, like a guard dog, ‘stop, heel’ (laughs) but you know it works for us.” (Joe)</i></p>	<p>Identifier: C2.24</p>
<p><i>“The best surface to walk on is a hard surface, smooth and hard” (Polly)</i></p>	<p>Identifier: PCA2.24</p>

Depth perception difficulties mean that Polly finds using stairs as a problem, particularly descending. She has a strategy for going downstairs safely but admits she still has a great fear of falling.

<p><i>“On the stairs, it’s the downs. I worry about not putting my foot on the stairs. What I do as well I sort of try and get to know the width what the stair is in case I go a little bit to the edge then I would think, oh no no and I would bring myself back while I’m holding on. But I’m careful very careful on the steps. Oh, I think it would be awful to fall down the steps you could do yourself a damage.” (Polly)</i></p>	<p>Identifier: PCA2.25</p>
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Joe has noted Polly's difficulties and describes trying to guide her safely if she needs to manoeuvre steps.

<p><i>“She has a problem with depth perception. ... even when she walks out of this house. I say, ‘no you go on the right the hand rail is there’. I say, ‘you’re at the top, go down one’, and she goes down one as long as each step is consistent, the same height. ... I say ‘kick with your toe, you’re up against a step’ so you know it’s a step then you go up. Of course, it’s coming down I always say, ‘You’re at the step, you’re coming down, just shuffle, right, now step down’, then its repetitive walking down.” (Joe)</i></p>	<p>Identifier: C2.25</p>
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Going shopping is also tricky as Polly’s visual difficulties cause her to bang into people. This has resulted in Joe stopping Polly shopping. They both recall the same incident.

<p><i>“we hold hands, or I push the trolley when we go shopping. I did bang the trolley once when we were in. That was the worst thing I should have done in my life. A big bang and I did knock this lady. I said, ‘oh I’m sorry’. She was ready to give me one and then she saw I was with someone. She said, ‘have you got something wrong’ and I said, ‘I’m sorry but I can’t see properly’” (Polly).</i></p>	<p>Identifier: PCA2.26</p>
<p><i>“Polly used to accompany me [shopping] and you know there is these little cart things. ... but it was bloody dangerous. I said you nearly ran that woman down.” (Joe)</i></p>	<p>Identifier: C2.26</p>

Comment: Diminished visual capacities particularly *impaired depth perception* is causing Polly difficulty in walking and coming down stairs. Joe describes her balance as also being a problem blaming it on her hip operation. Pavements are a cause for concern. Their uneven surfaces cause problems due to balance as well as her vision and perception problems. Polly describes her energy as poor and Joe explains she cannot walk far. This lack of exercise and resulting loss of muscle tone may also be exacerbating her poor balance and making her at higher risk of falling.

4.3.5.8.2 Getting Lost

Polly wandering out of the house causes Joe a good deal of worry because she will forget where she lives.

<i>“She forgets where she lives at times and I got her bracelet with her name and telephone numbers engraved on it. If somebody says, ‘where do you live?’ she forgets ... we had this last night. (Joe)</i>	Identifier: C2.27
<i>“I went to the Chemist and I got back and she’d gone and somebody had took her in up there and you know she just said she was going to the hairdressers for a chat ... and basically I had to sign for her and it was the girl next door’s ex-husband, he was driving one of these estate cars with ambulance on the side so he had all that where she says ‘I want to get away, I want to get away. There have been a few incidences.” (Joe)</i>	Identifier: C2.28

This sometimes proves dangerous. He describes Polly trying to leave the home and when stopping her, causing her an injury.

<i>“She was trying to get out the door once, I put me hand round and went like that and she had a watch and it scraped all her skin”. (Joe)</i>	Identifier: C2.29
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Polly, on the other hand, although admitting that Joe didn’t allow her to go out on her own, asserts that she didn’t go out alone, it was always with Joe.

<i>“Like Joe would say I’m not allowed to go out on my own and that but even just going down the garden” (Polly)</i>	Identifier: PCA2.27
<i>“I don’t go out on my own. I’d maybe go to a (long pause) oh there is nothing near, there’s nothing round me where I would go on my own. I have to be taken and brought back.” (Polly)</i>	Identifier: PCA2.28
<i>“I wouldn’t be silly enough to wander away. When we’re out I keep my eye on where Joe is all the time.” (Polly)</i>	Identifier: PCA2.29

Polly admits she sometimes gets lost in the house but claims that this is Joe's fault because he keeps moving the furniture.

<i>"You know I've gone into rooms and I've thought oh I can't see that right" (Polly)</i>	Identifier: PCA2.30
<i>"Joe is a mover of furniture and I am trying to say please don't move things around. I do get myself lost now and then but I do get myself out of it." (Polly)</i>	Identifier: PCA2.31

Joe describes a recent incident when Polly had not recognised her bedroom.

<i>"She's come in her bedroom confused. Even last night she said where's me bedroom. I say here's your bedroom. Even yesterday she said, 'where's my bedroom?', and then she'll go in there and say it's terrible, it's too small." (Joe)</i>	Identifier: C2.30
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Comment: Polly is displaying symptoms of *environmental agnosia* both inside and outside the home although Polly blames Joe for her difficulties inside the home saying he moves furniture around which confuses her.

4.3.5.8.3 Light and dark

Polly finds it difficult to navigate her environment in low light levels.

<i>"I don't like it when its dark (Polly)</i>	Identifier: PCA2.32
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Joe is aware of this and has tried to remedy the problem by using movement sensor lighting or physically being there to support her.

<i>"She does have problems with the dark. ... I must admit when we go and get the car I always leave the outside light on and I go and open the door and then I go back to her and then we come back and get in. ... She is very unsure because she can't see if it's dark." (Joe)</i>	Identifier: C2.31
<i>"... In the bedroom she has this light which is on all the time but as soon as she moves another one comes on so it's not as though we haven't attempted to find things to help her" (Joe)</i>	Identifier: C2.32

As well as low light levels, Polly also finds glare challenging. Joe has ensured that Polly has her eyes tested regularly and that she wears tinted glasses.

<i>I think glare is a real problem. I go every two years [the optician] but with Polly having eye problems, she goes every year and even got an ordinary pair of glasses they can tint down. She has them in her bedroom. Occasionally she has to put them on in the house because she says it really helps because it reduces glare but that's what we class as a smart pair. The one from the Blind Institute is what I leave in the car ... In the summer when we're in the car I say, 'put your glasses on' because it reduces glare." (Joe)</i>	Identifier: C2.33
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Comment: Polly has diminishing visual capacities of *sensitivity to glare* and *problems with low light levels*. Joe is aware of this ensuring she has the correct glasses and providing movement sensor lighting in her bedroom. Such visual problems also occur with glaucoma as well as PCA so the impact of having both conditions could be exacerbating her visual experiences.

4.3.5.8.4 Colour and Contrast

I ask Polly if she thinks certain colours help her to see better. She responds that she hadn't ever thought of colour helping her but after consideration says she enjoys looking at bright colours.

<i>"It varies a lot but not dark colours. I'm not interested in dark colours, but I like oranges and light fruity colours." (Polly)</i>	Identifier: PCA2.33
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Joe totally dismisses the notion that different colours or colour contrast could help Polly and appears to be distrustful of people telling what he should or shouldn't do regarding their home environment.

<p><i>“You know some people say I should have stripes on the doors. It would end up like kaleidoscope village. If Polly said, ‘oh I can see red over any other colour’, if you did everything in sodding red you are not going to notice any difference are you? I can’t really comprehend their thoughts cause I haven’t looked into it. The way they said it it’s a generic solution to people who can’t see very well in the home. There you go, slap this thing on and that will cure everything. It might in some cases but not for Polly and people tend to think that Oh yes, she’s got sight problems, oh do this, do this, but they don’t know how involved it is. I am not making it difficult, the fact is it is difficult.” (Joe)</i></p>	<p>Identifier: C2.34</p>
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Comment: Polly’s love of bright colours might be because she can see red and orange tones easier than other colours. Joe makes the point that if you did decorate in one colour everything would become invisible. He expresses the view that it is not him being difficult, he feels it is an impossibility to help Polly this way.

4.3.5.8.5 The use of devices to promote independence

Apart from cooking utensils for visually impaired people (which Joe did not let Polly use), the couple do not own any other accessible products that might help promote Polly’s independence. Joe feels they would be a waste of time because Polly constantly forgets how to use things.

<p><i>“We’ve even been to the blind institute and looked at all their things and even computers and you can pay a hell of a lot for a computer that can talk to you every time you press one of these keys, it confirms what you’ve pressed but unfortunately, if you showed her something today by tomorrow she’s forgotten it. Like on the remote for the telly.” (Joe)</i></p>	<p>Identifier: C2.35</p>
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Comment: Joe does not consider gadgets that help people with visual difficulties will be useful for Polly as he thinks she will just forget how to use them. The cost of them also seem to be deterrent although he has already stated they have several RNIB gadgets in the kitchen cupboard which Polly doesn’t use.

4.3.6 Support networks and their understanding of PCA

4.3.6.1 Family

Polly and Joe's only regular family member who helps them is their daughter who comes to see Polly every week to take her to an Alzheimer's club meeting. Polly describes this as the highlight of her week, not only because she is with her daughter, but because it allows her to socialise with others.

<i>"... my daughter comes every Tuesday. That's when we go Alzheimer's. Carpet bowls we do. ... Cause I do the [Alzheimer's] club and then the other part of the month we do another one. I went to that singing. I just join in everything" (Polly)</i>	Identifier: PCA2.34
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Joe advises that he has a sister but that they are not close, and they didn't have regular contact. A cousin of Polly's keeps in touch but only by telephone and she isn't really understanding of Polly's condition.

<i>"...oh, there's nothing wrong with you and I say, yes, cause I am talking to you on the telephone." (Polly)</i>	Identifier: PCA2.35
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4.3.6.2 Friends

Joe describes the couple as having few friends.

<i>"That's quite unfortunate really. We're not ones for keeping in touch. All those years [elsewhere] we didn't regularly write to folks. Friends from there, they're all scattered around. Most of the time, it's just the two of us." (Joe)</i>	Identifier: C2.36
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Joe admits that Polly enjoys the company of people but in the main there was only himself and their daughter most of the time.

<i>"she [daughter] is working so it's not as though you can grab people who walk by and say come and sit with the wife, you know. That's the situation." (Joe)</i>	Identifier: C2.37
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Joe describes a commemoration event a couple of years ago. Because of Polly's PCA, she had been unable to visually recognise people and he was angry because he felt they were ignored.

<i>"...and I took her down and of course she couldn't see them, and they'd say, 'oh is Polly, hiya', and you know she couldn't see them. She remembered their names. ... And you could see those people weren't enthusiastic to come and sit down with her and I saw that, and I thought it was a bit hurtful for Polly. And you could see on their faces, it was 'oh she doesn't recognise us' and they don't really want to get involved, they don't turn round, and say come and sit with me and let's have a chat. No, they don't (Joe)</i>	Identifier: C2.38
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4.3.6.3 Health and Social Care Professionals

Social services pay for Polly to have a support worker, eight hours per week and Polly enjoys her visits.

<i>"I do my timetables and I will go right through it for a week and then we'll go on to spellings and sentences and I'll sit, and I'll try to write. We were writing the other day. [She] tried to make me a plinth, a guide, you know. We do things like that and then we'll go out." (Polly)</i>	Identifier: PCA2.36
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Unfortunately, Joe does not like the woman.

<i>"I don't like the support worker, but Polly likes her." (Joe)</i>	Identifier: C2.39
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His relationship with the support worker deteriorated after she asked for double time while working on a bank holiday. He explains angrily:

<i>“I says just a minute, I checked up on the blooming description and everything and the rules and its says if public holidays fall on a normal working day normal pay is given, no extra time. Oh well I still want double. So I said well I’m not giving it you. ... I tell you she wouldn’t have stayed here if Polly didn’t like her, she’d be far gone. I don’t need that. I don’t need that at all. ... I find it quite frustrating cause she’s here to help Polly and you know Polly thinks the sun shines out of her backside, but it doesn’t, it doesn’t. Sorry I’ve rattled on.” (Joe)</i>	Identifier: C2.40
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Although, he admits he is constantly stressed and short-tempered, Joe has also refused the help of a cleaner:

<i>“a few weeks back one of these ladies [social worker] that come turned round and said ‘would you need a cleaner?’ I said, ‘I don’t want anyone coming in’. It’s one of those things on so and so is coming at this time. You feel as though you’re just living your life to be here whilst they come in.” (Joe)</i>	Identifier: C2.41
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Joe mentions that they were still waiting to be allocated a case worker, but this had been held up because the social worker they had first met had been off on extended sick leave. He finds the whole situation exasperating.

<i>“you know it’s quite frustrating. And that’s the same people who come in and say this this, this, this and if you want to have clarity on something oh no she’s not in the office.” (Joe)</i>	Identifier: C2.42
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He also talks scathingly about the respite care they had been offered and how it did not meet Polly’s needs.

<p><i>“In case you need respite ... there are two static caravans on the beach on the cliff top ... or this cottage ... So, I said can you send me particulars? So, I am reading about this cottage; it’s quite isolated, you can’t drive the car to it. You have to get out and walk on a path which is quite dark, there are no handrails and once you get into this house, it’s on different levels [no lift] and I said to Polly ‘Christ I bet they have an ambulance parked round the corner when anyone moves into this sodding place, its bloody ridiculous’. This house, this was offered to us because it’s for people like us. There’s a wet room in there which she can possibly use. So, this house is adapted. That bloody house, I said it’s an accident waiting to happen, so we never pursued it” (Joe)</i></p>	<p>Identifier: C2.43</p>
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Joe also explains that Polly had kept having blood tests and they never had any feedback. When he enquired, he found the surgery had put her results in another person’s file who had a similar name. This had also resulted in them nearly being given the wrong medication at the pharmacist.

<p><i>“I went to the chemist for summat and he said, ‘I’ve got a prescription for your wife’. I signed for it and as I’m signing ... I just happened to look ... and it was a different address. ... I was getting annoyed.” (Joe)</i></p>	<p>Identifier: C2.44</p>
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He is also angry about getting into trouble because he had not taken Polly for eye screening after he was told it wasn’t necessary by the doctor.

<p><i>“I mentioned it [eye screening] to the doctor last time and I said do you know, a few years ago my wife was diagnosed with PCA? They said there was no need for her to go ... and then last year ... they got on the phone and threatened us with everybody. You must do this, you can’t do that. I said my wife’s diabetic type 2, she’s got glaucoma, she’s partially sighted, she’s got PCA. Do you know what PCA is? Yes, I know what PCA is?. Oh honestly and as soon as you go into the system and you say she has PCA oh we don’t know about that. I’m not saying because she’s got PCA she must not have all these tests. I’m not saying that, but it seems as though it is putting her through uncomfortable procedures when it is possibly not necessary.” (Joe)</i></p>	<p>Identifier: C2.45</p>
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Joe has also found that health and social care professionals often do not understand what PCA is and do not respond either appropriately or sympathetically.

<p><i>“You know like when you get flu you get heady then you start sneezing, then you blow your nose. You go through a course, but I can’t say PCA is like that, it’s so variable and I say variable cause it’s a fact. ... at times Polly’s got frustrated but I got bloody frustrated because you can tell people and you can see its going over their bloody heads. They are not listening to you and many times I feel like saying ‘Hey will you just put your pen down, sit on your backside and listen to what we have to say’. And I found it very frustrating.” (Joe)</i></p>	<p>Identifier: C2.46</p>
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Comment: *The couple have a poor social network. This is affecting their relationship because they are together most of the time and don’t get a break from each other. Joe says he is stressed but appears to refuse any help offered by social services that would support them. Social care professionals appear not to understand the symptoms of PCA as the respite care accommodation offered was not suitable for Polly’s specific visual and perceptual needs.*

4.3.7 Changing roles within the couple’s relationship

Both individuals have found the changes in their respective roles difficult and this appears to be having a negative effect on their relationship.

Polly is resentful:

<p><i>“My biggest problem is my husband. ... Now Joe’s took the house over, I hate it. He’s took nearly everything off me.” (Polly)</i></p>	<p>Identifier: PCA2.37</p>
<p><i>“I do not love him 100% because of the years go on. He’s very very bossy and I get on his nerves when I can’t do it. This morning he was going to start on something and I stopped him. He started last night, I said for goodness sake Joe, just give me a bit of leeway.” (Polly)</i></p>	<p>Identifier: PCA2.38</p>
<p><i>“Yes, a lot has been taken away from me, as far as Joe. Please don’t say anything.” (Polly)</i></p>	<p>Identifier: PCA2.39</p>

Joe, on the other hand, appears to be becoming overwhelmed by his responsibilities.

<p><i>“It’s getting worse. ... I do the washing, I do the ironing, there are times I look and say, ‘oh that’s dusty I better dust’. It’s not as though I’ve got a weekly ritual. I like to think I am proactive rather than reactive and I do the cooking but I do get tired sometimes.” (Joe)</i></p>	<p>Identifier: C2.47</p>
<p><i>“I do the shopping now. I write the list. Now I wait until someone is in the house and then I shoot off. ... we used to get a barrow and I’d say you push it and I used to be at the front guiding it but then she was stood in the centre of an aisle, oh I’m going back about four or five months and I’d gone on and I said, ‘what’s up?’ and she was sobbing her heart out because she can’t look whether it was raspberry or strawberry jam, she can’t differentiate. She can’t do this, and she can’t do that, so I sez, ‘no I’ll do the shopping’ so I go and do the shopping.” (Joe)</i></p>	<p>Identifier: C2.48</p>
<p><i>“I have a lot of drugs to manage each day and each week. Well it helps that I’m a methodical person. What I go and do, these are the morning ones, all the way up there. Then I put the teatime ones in there and the night time ones are in her bedroom” (Joe)</i></p>	<p>Identifier: C2.49</p>

4.3.7.1 Caregiver Stressors

It hadn’t helped matters when Polly stated she wanted a divorce!

<p><i>“I said. ‘we can’t get divorced we’re bloody 70. I haven’t got sodding money to get divorced’.” (Joe)</i></p>	<p>Identifier: C2.50</p>
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Joe explains that a few months ago, Polly had gone to her GP and accused Joe of hitting her. The doctors had reassured Joe that they believed him, but social services arrived at the house and took Polly away to a Care Home.

<i>“I feel as though I am coping but you know when she takes off and does this, I think, ‘oh shit.’ You feel it can’t get any worse cause you feel you are at the limit already.” (Joe)</i>	Identifier: C2.51
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Within 24 hours, Joe was asked to take Polly home. When they were leaving it was suggested that Polly came to them one day a week. Despite being so stressed, Joe again refused their help.

<i>“... we got on the door step and there was two of the workers and one turned round and said ‘you know if you want to bring Polly, it only costs £20 for during the day’. I nearly turned round and said are you on commission or summat?. £20 to look after me wife! And then Polly said, ‘I don’t want to go there anymore’. I said, ‘Well I didn’t take you there. It’s just events that had happened cause you’d gone bursting in, it activates something else naturally”. (Joe)</i>	Identifier: C2.52
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Comment: Polly is displaying some challenging negative behaviours against Joe but is this due to her frustrations and boredom of her situation? The fact that she had asked for a divorce indicates the situation is nearing breaking point. Joe appears to be becoming completely overwhelmed by his situation and such continued stress may impact on his own health. Even though he admits to being stressed he is still refusing support from social care. Is this due to the fear of the cost of care rather than not being willing to accept it or a reluctant to lose an element of control over the way he cares for Polly?

4.3.8 Wanting to protect the person with PCA

It is clear Joe is doing his best to protect Polly from hurting herself.

<i>“I don’t let her do too much in case she cuts herself or burns herself. I think well it’s not worth it and that basically is what it is so what do you do” (Joe)</i>	Identifier: C2.53
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But sometimes this protective streak seems to go a little too far. Polly describes a holiday when Joe locked her in her room each night so that she wouldn’t wander and get herself lost.

<i>“... there was this trip. ... Joe said I think we need separate rooms and what we did was we have separate rooms. We had one that had a toilet and shower in and one where I could close the door and the window and then Joe had a key to lock me in. I didn’t like the idea but when we went to bed he locked me in. So, he would lock me in and then he would go to bed. ... and when the holiday was nearly at an end a man said to me did you find your husband the other night? I said pardon? He said you were walking round and round say Joe, Joe. That why we made him lock. I knew it was a bit of a gamble to lock but it was that or no holiday. I would have gone out, definitely. ... But he didn’t like it. He was embarrassed, embarrassed. (Polly)</i>	Identifier: PCA2.40
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Polly agrees that she needs protecting from hurting herself or getting lost but during our conversation, the frustration of losing her identity and independence is clear:

<i>“What it does it make’s people that’s with you behave so you’re a little girl, you get treat like a little bairn. And I try and show them I can do it. Just watch me when I come to big things like I’m going to cross that. Keep your eye on me but please don’t Joe. I say I can see, I can see the things in the floor. I miss a few but not when they do it sometimes I think Oh gosh!” (Polly)</i>	Identifier: PCA2.37
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Comment: *The way the couple handled their holiday was not ethically acceptable. If Polly had needed to evacuate her room due to fire the results could have been catastrophic. Joe had tried to manage the situation himself and did not think to ask for support from others be it social care or even the hotel staff. Polly's perceives she is being treated like a child because she is continually told not to do things.*

4.3.9 The emotional impact of PCA

Polly expresses the sadness she feels losing her identity and role in life.

<p><i>“Sad, happy, stressed. You feel a bit of all them things. You can’t help it, it’s a part of your life. Everyday life. You can’t knock that part out for good. No, I miss, I miss being Polly the housewife, the shopper. I’m not giving in.” (Polly)</i></p>	<p>Identifier: PCA2.38</p>
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<p><i>“I wouldn’t use lifts, not on my own. Somebody might rape me, rob me, bang me, hurt me and I know I’m awful, but I do worry about people doing things to you.” (Polly)</i></p>	<p>Identifier: PCA2.41</p>
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Joe tries to understand Polly’s feelings

<p><i>“There have been odd occasions when she has been crying and I say, ‘what’s up?’. I’ve lost you, I’ve lost you. Possibly feeling so isolated or something”. (Joe)</i></p>	<p>Identifier: C2.54</p>
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But expresses the situation in a bleak manner

<p><i>“I tend to think, is there any hope?” (Joe)</i></p>	<p>Identifier: C2.55</p>
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Comment: *The couple are both displaying very negative emotions including high levels of stress and intense unhappiness. Polly refers to ‘crying alone’ on several occasions during the interview. Joe is trying to understand Polly’s feelings but his own feelings of helplessness regarding the situation seem to be becoming overwhelming.*

4.3.10 Facing the future

Neither of the couple are positive about future. Although Polly says she is going to fight to be around for her daughter, her words show a more pessimist outlook:

<i>“I thought, ‘I’m not dying’. So, then you think, ‘thank God, I’m not dying so what you’ve got you’ve got to make it do and enjoy it as much as you can.” (Polly)</i>	Identifier: PCA2.42
<i>“I want to be positive, but I’ve had so much bad luck in my life, you can’t. You can say I’m going to try and try but what God gives you, you can’t fight, you know. But I’m going to try. I want to live as long as I can. I want to be with my daughter and live as happy life as we can. I just wish, I want it to be nice. I like life, I like life. I like doing things. I like doing unusual things.” (Polly)</i>	Identifier: PCA2.43

When I ask Joe about if he thought about the future, the angry man changes into a very tearful individual explaining he has been given very negative news about Polly’s future, and just how hopeless he feels the whole situation is.

<i>“... you know it was said to me ‘oh I wouldn’t stand for that, I’d bugger off, but it’s alright saying bugger off but it’s not as though we are new to it. No, I have not even looked at it as an option”. (Joe)</i>	Identifier: C2.56
<i>I’m, positive to be negative. I know that sounds contradictory, but the doctor said to me, ‘Joe, you’ve lost Polly, you won’t get Polly back. ... it’s been a gradual progression. It’s just like somebody chipping away. Anyway, they said ‘you’ve lost Polly, you won’t ever get her back. ‘(gets very upset).’ (Joe)</i>	Identifier: C2.57

Comment: Polly’s feelings about the future swing from despair to determination to carry on for as long as possible. Joe is in tears at this point, very emotional indeed. He is getting very negative opinions from others, both friends and health professionals, and this is making he even more desperate. He has no one to talk to who understands his situation or can encourage and support him practically and/or emotionally.

4.3.11 Summary of Polly and Joe's story

Polly is living with several long-term conditions as outlined in [Table 17](#) below:

Table 17: Polly's long-term conditions

●	Diabetes
●	Emphysema
●	Glaucoma
●	High blood pressure
●	Psoriasis

She has also had a full hip replacement and cataract surgery

[Table 18](#) outlines Polly's PCA symptoms:

Table 18: Polly's PCA symptoms

●	Alexia
●	Anomia
●	Apraxia
●	Associative Agnosia
●	Dressing Apraxia
●	environmental agnosia
●	impaired depth perception
●	Increased sensitivity to glare
●	Prosopagnosia
●	Simultanagnosia

Polly had had poor vision for many years and so it is difficult to gauge when her PCA symptoms began. Her glaucoma seems to have masked the real reason for her vision and perception problems and so resulted in delays in diagnosis.

Even though she knew her vision was poor, Polly continued to drive until she felt it was dangerous to do so.

The couple did not receive emotional support or a good explanation about PCA once Polly was diagnosed and her personhood was undermined by a psychiatrist who spoke in a condescending and inhumane manner.

Polly has lost the ability to read, write and understand financial matters. She also has difficulty with personal care, dressing herself and feeding herself but her appearance is still an important element of her personality. She misses her role as housewife and even though they have many RNIB products which might help her to continue to undertake some tasks, Joe prevents her, and this lack of team work is causing Polly deep resentment against him.

Polly is aware she is not perceiving people, objects or scenes correctly. She sometimes does not recognise Joe or her home and gets lost easily both in inside and outside environments.

Polly has low energy and has balance problems. Impaired depth perception particularly cause difficulties in descending stairs or walking on uneven surfaces making her at high risk of falling. This is also exacerbated by her increased sensitivity to glare.

Gadgets or colours that might help Polly to see or manipulate her environment better are dismissed by Joe who appears to want full control of the household and what Polly can do.

The couple have a poor social network. This is having a toxic effect on their relationship and Polly has asked for a divorce which Joe has dismissed stating they are too old to do such a thing. Joe is refusing most support offered by social services except for a support worker who comes in a couple of days a week. Respite care holiday accommodation was offered but it was not suitable

The couple of both displaying high levels of stress and intense unhappiness regarding their circumstances. Both report feelings of helplessness.

Polly's thoughts about the future swing from despair to determination to carry on for as long as possible. Joe is getting negative opinions from others, both friends and health professionals, and he is becoming emotionally overwhelmed. He appears to have no one to talk to who understands his situation or can encourage and support him. The wellbeing of both individuals is poor.

4.4 Case Study Three: Brian and Barbara's story

Brian and Barbara were interviewed separately in their own home in different rooms to ensure complete privacy. Each interview lasted one hour, was tape recorded and fully transcribed.

4.4.1 Setting the Scene

Brian is a 61-year-old gentleman who has had a diagnosis of AD with PCA for 18 months. He appears nervous during the interview and often has trouble expressing himself, with difficulties in word-finding. He has been married to *Barbara* for 38 years and they have one daughter who was living away from home, studying at University. The couple have both lived in *Sheffield*, in the same house, all their married lives. Brian left school at the age of 15 years. He is very interested in music and got his first job in a local Hi Fi shop eventually working up to being store manager. His main hobby was cars and he enjoyed rally driving in his spare time. When the store closed, Brian was offered another job immediately installing and selling similar products but started to find it difficult to cope with the pressure of work. It was agreed he should reduce his working hours to part-time. He finally finished employment due to ill health when he was 58 years old. Barbara gained a degree in pharmacy and worked as a pharmacist until retiring at 55-years-old to care for Brian.

The couple have lived in the same semi-detached house for 38 years in a quiet suburban area. The décor is tidy with plain carpets and Barbara has equipped movement sensor lights in hallways and stairs to help Brian find his way around the property. The downstairs consists of a large lounge and small kitchen area which leads onto a bright conservatory where the couple spend much of their time. They have a small garden designed for easy maintenance. The upstairs consists of three bedrooms and bathroom which contains a bath and shower area.

4.4.2 Realising something was wrong

Barbara recalls that Brian's symptoms had begun about six or seven years previously. They had both had a bad year: Brian had lost his job, his mother died, and his best friend's wife had died of cancer. Brian secured a new job, but Barbara started to notice he was acting strangely:

<i>"I know it's a typical joke that you know man goes to a supermarket and can't see a tin of soup in front of him, but he was really like that. He couldn't see a mug on the table, but he could see the crumbs. It was just weird. How could he see the crumbs and not the mug?" (Barbara)</i>	Identifier C3.1
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<i>"He seemed in a mad rush all the time but doing weird things like (sighs, pauses) he'd get in the car and drive off without putting his seat belt on and then try and put it on when driving. ... His driving was getting more and more peculiar. Swerving round things that weren't there and veering to the left a lot. He might pull out round a dark patch of tarmac. He said he needed to do it." (Barbara)</i>	Identifier C3.2
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Brian describes how he had felt:

<i>"This sounds strange, really strange. When I was driving (pauses) the steering wheel was going like that (makes side to side moves) ... moving on its own to the left. ... I wasn't, I didn't even know. I wasn't aware I was steering. I thought I was going straight. Barbara said, 'watch what you're doing.' The other thing is there were things on the road ... like shadows, and I had to drive round them. I thought, 'I'm going to crash or there's somebody in front'. It's just like a ghost or something." (Brian)</i>	Identifier PCA3.1
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Barbara says she suggested he had his eyes tested.

<i>"It felt like he was picking fault with things, complaining there was a bit of mess there, crumbs or it might be water spots on the floor that he could see but not the thing he was looking for. I thought he was just going peculiar ... and being awkward" (Barbara)</i>	Identifier C3.3
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Comment: Visual perception problems are described by both Brian and Barbara. Brian is experiencing hallucinations – thinking tarmac is a solid object or seeing someone in front of the car. He was also missing part of a visual scene (*simultanagnosia*) such as spotting crumbs on a table but not a mug.

4.4.3 Journey to diagnosis

Brian went to his local optician and had an eye test.

<i>“They gave him new glasses. Brian said they didn’t help but the optician said they were fine for his eyes.” (Barbara)</i>	Identifier C3.4
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Things did not improve so the couple next visited their GP.

<i>“He thought it was anxiety which seemed quite likely after the bad year we had had and starting a new job where there were a lot of younger people who knew everything.” (Barbara)</i>	Identifier C3.5
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Unfortunately, the situation began to get worse.

<i>“They were sending him out to people’s houses to install electronic equipment and I found he was really struggling with maps. He couldn’t read maps and he was getting lost.” (Barbara)</i>	Identifier C3.6
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Brian would ring Barbara and ask her where he was. She started using Google maps to help him find his way.

<i>“He was getting very stressed and worried ... getting to people’s houses in a real mess. He couldn’t sort out what the problem was, and it even ended up that he couldn’t put a three-pin plug on and he used to install really complicated Hi Fi.” (Barbara)</i>	Identifier C3.7
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Barbara felt that the visual problems were more than just anxiety and they went back to the GP.

<i>“We just went to different therapists and counsellors and doctors and opticians and it just went on and on.” (Barbara)</i>	Identifier C3.8
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Barbara’s sister, who is an occupational therapist, asked if the specialists had ruled out PCA. At the same time, Brian’s sister contacted them to say she had read an article in a national newspaper written by a middle-aged woman with PCA and that nobody had recognised it and how dreadful it was to cope with because nobody understood.

Barbara acted:

<i>“I wrote a very tactful letter to the neurologist saying my sister had seen him in a social setting and realised he had a visual impairment and had they considered PCA?” (Barbara)</i>	Identifier C3.9
<i>“He’d had scans, but they couldn’t see anything on the scans, they said. I had a health insurance and sent the scans to Boston University in America and they said they could see some atrophy on the scans.” (Barbara)</i>	Identifier C3.10

The doctors in UK disagreed, advising that they could find no evidence of change in his scans. However, the specialist accepted her argument.

Brian reduced his hours at work, finally stopping employment two years ago after he was diagnosed in 2015 with AD with an emphasis on PCA. He undertakes a medical check-up once a year.

<i>“.. he has a medication review once a year ... just medication and they do a mini mental state thing, but he can’t do it anyhow you know counting backward from 100 in 7s and draw a clock. He just can’t do it. ... His memory is no worse than mine ... his vision and lack of confidence are the main problems.” (Barbara).</i>	Identifier C3.11
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Brian's medications include donepezil and memantine for dementia; venlafaxine for depression and anxiety and levothyroxine for an underactive thyroid.

Brian explains that he stopped driving completely after a frightening driving incident.

<i>"I started getting beeped at and I was going round a roundabout, going home. I got lost on the roundabout and I ended up going the wrong way. ... It was awful. I didn't know what I was doing. (Sighs). Anyway, I didn't crash but I hit somebody's mirror, and I'd never done that before, never. And his glass shattered, and I just drove off straight away and I didn't wait to be (pause) I was scared. There was a terrific bang." (Brian)</i>	Identifier PCA3.2
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Comment: *Initially, the optician and the GP do not suspect any form of dementia. Because he has had a stressful year, the first diagnosis is anxiety. Brian starts to get lost going to client houses (*environmental agnosia*) which increases his anxiety levels. An occupational therapist rather than a doctor is the first person to realise that Brian might have PCA but brain scans are inconclusive. He is finally diagnosed with AD with PCA. Interestingly Brian is still driving at this point and almost has a serious accident when he becomes disorientated on a roundabout.*

4.4.4 Reactions to knowing

When asked if he could explain what PCA was, Brian responds:

<i>"It's a funny sensation. Things started to happen with my brain. I assume it's my brain." (Brian)</i>	Identifier PCA3.3
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But for him, the worse thing about being diagnosed appears to be having to give up driving. The loss of his favourite pastime is still very painful to talk about.

<p><i>“I loved my cars and that was my passion. ... keeping it clean and hoovered. ... I loved my car and I loved driving it. Being in control of a car, driven well, is something else and it came on so quickly.” (Brian)</i></p>	<p>Identifier PCA3.4</p>
<p><i>“I can drive. I know I can drive but I’m not allowed to drive, and I said, ‘well what happens if I don’t give my licence up?’ And she said, ‘the police will come and take your licence off you’.” (Brian)</i></p>	<p>Identifier PCA3.5</p>
<p><i>“I used to do rallying so it’s very high on my must haves and every time you turn on the telly or the radio they are trying to sell you a nice car. That’s the main thing I’ve lost. ... And it came on so quickly. ... I’ve been trying to see if I can have some (very long pause) some lessons. But I don’t need lessons. I know how to drive. I can move a vehicle. But they won’t let me.” and this [PCA], that’s the worst thing that could have happened to me” (Brian)</i></p>	<p>Identifier PCA3.6</p>

Barbara is pragmatic in her response to the diagnosis.

<p><i>“He gets lonely. Once I realised that what he’d got was only going to get worse, I felt it made even more sense to retire and be at home and go off doing things together while we can. Which is what we are doing.” (Barbara)</i></p>	<p>Identifier C3.12</p>
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Comment: Brian expresses a great sense of loss and sadness as his favourite hobby – driving – has been taken away from him because of PCA. Barbara’s career ends as she takes early retirement to look after him.

4.4.5 Day to Day challenges

Brian is finding activities of daily living becoming harder to do and relies a lot on Barbara to enable him to carry on living as well as possible.

4.4.5.1 Personal Care

Brian can still accomplish most of his personal care himself. Difficulties arise in trying to manipulate equipment such as his shaver or nail clippers.

<p><i>“I have to remind him when he needs a shave. ... He only shaves about every three to four days cause he’s got a beard. I’ve suggested he shaves with his eyes shut but he hasn’t tried it yet. But I think that might help. A couple of weeks ago he was shaving his beard and he shaved it all off on one side, so I had to shave the other side to make it match.” (Barbara)</i></p>	<p>Identifier C3.13</p>
<p><i>“he can’t cut his fingernails. ... He really struggles with that and his toenails. I take him to the chiropodist ... and they cut his nails.” (Barbara)</i></p>	<p>Identifier C3.14</p>

Barbara normally sets the shower going for him to get the temperature right as Brian finds it difficult to use switches. He is unable to identify washing products by the writing on the packaging, but he can differentiate using colour.

<p><i>“I have to tell him like the grey one’s for your hair and the brown one’s for your body. I go by the colour. ... He’s fine with that.” (Barbara)</i></p>	<p>Identifier C3.15</p>
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Comment: Brian’s motor skills (*apraxia*) are now compromised and he needs help with personal care. He is experiencing alexia and cannot read labels but can still identify products by their colour.

4.4.5.2 Dressing

Although he can still dress himself, this activity is becoming increasingly difficult and slow. Brian has trouble in finding his clothes explaining that he can never locate them in cupboards. He is sensitive to the fact that Barbara has suggested putting patches on the back of garments to help him find them. To Brian this put more emphasis on the fact that he is ill.

<p><i>“I don’t do cupboards. I can never find anything in a cupboard. Barbara says, ‘there it is there’. She is threatening to sew patches on the back. I don’t want her to do that. No, it’s like’ oh it’s come to this has it.” (Brian)</i></p>	<p>Identifier PCA3.7</p>
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Barbara lays out the clothes he is to wear in the same place each day, but Brian still finds it difficult to locate them.

<i>“He usually knows which clothes he wants but he can’t find them. He’s got a blanket box at the end of the bed and his clothes are normally on that, but he’ll say he can’t find them.” (Barbara)</i>	Identifier C3.16
<i>“He has a lot of problems getting dressed. He’ll get things inside out, upside down. He gets the sleeves pulled inside and sometimes he’ll bring something to me and it’s like he’s been knitting with it. One sleeve inside the other and then all twisted round and he just can’t sort it out.” (Barbara)</i>	Identifier C3.17

Brian admits that he dresses slowly and prefers Barbara to be there for support.

<i>“I normally sort of dress slowly and I’ll dress while Barbara is there”. (Brian)</i>	Identifier PCA3.8
<i>“Sometimes I find it difficult to put my trousers on and shoe laces and coat. It’s a nice coat, it nice and warm and it looks nice but it’s difficult to put on at times. ... if I’m stressed I daren’t do it. It’s got one of these zips which is three together like that and I can’t do it but on my own because I can’t see it so when you are putting it on, it’s got three zips like that. Barbara can do it for me, whoosh, whoosh. She can do it easy. Now the thing is Barbara says if you carry on like this you’re going to break the zip and I don’t want to break the zip so most time, definitely when I’m with Barbara, she will zip me up so that’s OK.” (Brian)</i>	Identifier PCA3.9

Comment: Brian is experiencing *dressing apraxia*. Difficulties in locating items such as clothing suggests *simultanagnosia*.

4.4.5.3 Food Preparation

Barbara encourages him to prepare simple meals even though it might end up a messy affair. She has used bump stickers supplied by RNIB to help identify the ring he needs to switch on the hob. They are using a gas hob, but Barbara doesn’t express any concerns that Brian might burn himself.

<p><i>“He doesn’t do any baking, never did. He can make soup but that’s his limit. He was getting into a bit of a mess over which ring was the right one, which knob related to which ring on the gas hob, so I’ve put orangey red dot on the knob for the ring that he always likes using and that solved it.” (Barbara)</i></p>	<p>Identifier C3.18</p>
<p><i>“He misses the bowl ... he holds the pan not over the bowl and also its lack of confidence. He pours it very, very slowly, so it tends to dribble down the sides of the pan and just goes all over the place.” (Barbara)</i></p>	<p>Identifier C3.19</p>

Brian voices concern that recently he has been finding this more difficult.

<p><i>“I will normally put a can of soup on and cook it. Yes, I can do that. ... Now just lately I’ve been, er, (very long pause) oh this is it I was perfectly happy pouring the soup out for one. For one person. Which is easier. You know the bigger the things are more difficult. So, I won’t do that. I cut myself off. ... Sometimes I make a mess of it and it might be all over the hob. Not the hob, the work surface. So, it’s not the end of the world but it does, it does annoy me. Again, if I’m stressed, I’m more likely to make a mistake and I can’t do more than three, can’t do more than three.” (Brian)</i></p>	<p>Identifier PCA3.10</p>
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Barbara also mentions that he can only manage two cups or two bowls of anything. She feels that after that he suffers from information overload which causes him to panic.

<p><i>“He makes cups of tea for just me and him, just two cups and he usually manages that. If there is three he ends up with two tea bags in one cup and none in the other or he pours the kettle into the sugar bowl. He just gets in a muddle ... its information overload. He can manage two but then it all goes wrong.” (Barbara)</i></p>	<p>Identifier C3.20</p>
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However, Barbara keeps encouraging him to carry on his housework activities even though it takes a long time or isn’t quite properly achieved, or he sometimes finds using the equipment difficult.

<p><i>“He occasionally has trouble with the kitchen taps. He said there was no hot water, but he was running the cold one and about two years ago he was complaining the kettle didn’t work but he was turning on the toaster and that’s why.” (Barbara)</i></p>	<p>Identifier C3.21</p>
<p><i>“He always washes the dishes and he still does now. It takes him ages, but he does do it. We’ve got a dishwasher, but he doesn’t want to use it. He always said he didn’t think it did as good a job as he did, which I probably agree with. But now I think he’s scared of it and doesn’t know how to load it, doesn’t know how to use it.” (Barbara)</i></p>	<p>Identifier C3.22</p>
<p><i>He hoovers. He gets in a tangle with the cable so that’s why we have got the cordless one ... he’s starting to like that, but I’ve noticed he does one bit and there would be all bits and mess in all other places. He’d only done one bit. I think he was just focusing on one bit and then forgot about the rest of it. (Barbara)</i></p>	<p>Identifier C3.23</p>
<p><i>There would be a mark on the worktop and he’d be rubbing next to it and saying the mark won’t come off. I said, ‘well you’re not rubbing the mark, you’re rubbing next to it’ but even that’s (pause) I think as he is understanding what the problems are, he’s learning to cope with it. (Barbara)</i></p>	<p>Identifier C3.24</p>

Comment: Brian is still able to undertake simple household tasks and Barbara is assisting him by thinking of ways to help. It is interesting to note that Brian can only cope with two things at a time. Trying to do too much at once can lead to disorientation and confusion in people living with dementia. Barbara refers to this as ‘information overload’.

4.4.5.4 Eating and drinking

Brian can still use cutlery but admits he is not as capable as he used to be.

<p><i>“I am not as accurate as I used to be eating but yes, I can hold my own.” (Brian)</i></p>	<p>Identifier PCA3.11</p>
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Barbara has bought some new place mats and now realises they have a busy flower pattern. She thinks that Brian may get confused using them but to her surprise, at the moment he has no problem using them.

<i>“I hadn’t realised how busy the pattern was on the mats. I thought this is all wrong, he won’t be able to see anything on the mats. but he really likes them because they are really flat so if you put a cup of tea on it, it feels really positive. It’s nice and smooth and so you can clean old food off it. And so far as the pattern on them, it doesn’t seem to be a problem which has sort of surprised me.” (Barbara)</i>	Identifier C3.25
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Comment: Brian is still able to use cutlery and crockery when eating and patterns do not seem to confuse him.

4.4.5.5 Reading, writing and numeracy

Brian now has great difficulty in reading and writing. He has been an avid book reader but now struggles. Brian seemed not to understand why he is having problems.

<i>“I was always reading. I start the book cause I’ve always been a reader but it’s, it’s, it’s, (long pause) I don’t understand. I haven’t done it for a while, (long pause) I’ve still got a book upstairs in the toilet (laughs)” (Brian)</i>	Identifier PCA3.12
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However, Barbara describes the difficulties Brian has spoken of when trying to read.

<i>“He used to read books. That’s something he’s stopped doing. ... he can’t concentrate ... the letters move. The lines jump up and down. He gets to the end of a line and then he doesn’t know which line to go to next. He’s backward and forwards doing the same line again and again and again and then he says the story doesn’t make sense. To start with I would get him a book, or he would get a book from somewhere and he would say ‘oh that book’s rubbish, the story doesn’t make sense’, and the same thing keeps happening again and again.” (Barbara)</i>	Identifier C3.26
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They have tried audiobooks, but this has not been successful.

<p><i>“Barbara said she’s got one of these talking book things. So, she actually bought the, the thing and she said do that cause I was enjoying Alan Titchmarsh. I couldn’t finish it. Couldn’t, couldn’t.” (Brian).</i></p>	<p>Identifier PCA3.13</p>
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Barbara is disappointed her idea hasn’t worked asserting that she thinks it may be Brian’s lack of concentration that is to blame.

<p><i>“He can’t concentrate. He’s tried audio books, but he can’t follow the train of it at all. Yes, cause I thought audio books would be something useful but no.” (Barbara)</i></p>	<p>Identifier C3.27</p>
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Brian’s writing had started to deteriorate about two years previously.

Barbara remembers him getting into a panic over writing Christmas cards, something he always used to do.

<p><i>“He sort of forgets how to form a letter so doing Christmas cards he gets in a real flap about that. In fact, two years ago, we ended up with so many envelopes thrown around. He was writing them with the address squeezed up in the top corner and I kept saying the address is meant to be in the middle, so he said, ‘OK I’ll do another one’ and then squeeze the address in the top corner again and we just went on and on. Cause I didn’t know what was wrong with him then. I just thought he was being silly.” (Barbara)</i></p>	<p>Identifier C3.28</p>
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When talking to Brian about this, he appears quite distressed. Barbara has suggested he try a children’s writing book to help him and he doesn’t want to do this.

<p><i>“... even the cards there (points to Christmas cards). I can’t even write the cards. I used to do all the Christmas cards. I can’t now.” (Brian)</i></p>	<p>Identifier PCA3.14</p>
<p><i>“His signature has gone very strange. I thought of paper but with wide apart lines ... I was thinking of primary school learning to write things where they’d got the letters</i></p>	<p>Identifier C3.29</p>

<i>like dotted lines to practice forming the letters, but he doesn't seem very keen about that." (Barbara)</i>	
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Using a telephone is also becoming problematic. Barbara has tried different ways of helping him see the telephone numbers when he wants to ring somebody, but this has not been particularly successful, with Brian resorting to asking Barbara to dial for him.

<i>"He really struggles with a telephone. I've got a laminated card with main phone numbers on, written big with pink stripes across so he's got phone numbers if he wants to ring anybody. ... He can just about deal with two digits at a time ... normally he brings the card to me and asks me to read the number out for him. .. I read the numbers in twos because that's all he can really manage but he often goes wrong. Maybe he'll take ages and press one number on the phone and spend ages looking for the next one and then he might press the wrong one and have to start again. He does get into a bit of a panic." (Barbara)</i>	Identifier C3.30
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Fortunately, an easy to use mobile phone with only two buttons is proving successful and Brian carries this around all the time for reassurance.

<i>"His mobile has two buttons across the top and it's me and our daughter and he can work that. ... He takes that phone when he goes to the toilet and everywhere, so he can get me." (Barbara)</i>	Identifier C3.31
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Brian can no longer understand money which causes him distress and he worries about appearing foolish.

<i>"I can't do money. This is another thing. First of all, I can't (pause) this is a stress thing. If I've got to pay with money. I get all agitated and then I make a mistake, or I show up myself. (Brian)</i>	Identifier PCA3.15
<i>"I don't like shopping now because it involves buying things and doing things which makes me nervous. So I don't do much shopping (Brian)</i>	Identifier PCA3.16

However, a tip for a member of the PCA support group to use a contactless debit card is proving a success and has really boosted Brian's self-esteem.

<i>"Yesterday I bought a Costa. It's contactless, yes. It made me feel very good. Very good. ... I've done it twice now." (Brian)</i>	Identifier PCA3.17
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Comment: Brian is experiencing *alexia, agraphia* and *acalculia*. Audio books do not seem to be successful due to his inability to concentrate. A successful tactic has been the use of a contactless credit card. Being able to buy a coffee without stress has made Brian's self-esteem rise. Barbara got this tip from a member of the London PCA support group that they had been able to attend.

4.4.5.6 Problems with visual perception

Visual perception problems were one of the first difficulties that Brian has experienced.

<i>"...there were things on the road ... like shadows, and I had to drive round them. I thought, 'I'm going to crash or there's somebody in front'. It's just like a ghost or something." (Brian)</i>	Identifier PCA3.18
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Although, he doesn't drive anymore, perception problems still cause the couple distress. Brian has trouble making sense of anything that is novel. For instance, the couple have got a new bed with new pillows, but Brian is finding it hard to get used to them.

<i>"We got a new bed. We got new pillows, quite thick fat pillows and he was lying halfway down the bed and he couldn't make sense of these new fat pillows. I said, 'just have one of them' and he said, 'oh I always have two pillows'. But they were great big fat pillows, so his head was half way down the bed and his feet were over the end. He said this was no good we need a new bed. It was really difficult about then." (Barbara)</i>	Identifier C3.32
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The quilt is also proving another difficulty. Although king-sized, Brian insists it is not big enough.

<i>“He’d be turning the quilt round on the bed and saying the quilt is not big enough and he’d just have a little corner of it over him. ... so, we ended up with a normal king-size bed with a king-size quilt. I went to sleep on top of the quilt to hold it and he sleeps underneath it and I have another small quilt on top of me to keep me warm and that seems to work. So, we are in the same bed though there is a quilt between us but at least it stays in the right place now.” (Barbara)</i>	Identifier C3.33
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Brian describes having a problem getting in and out of toilets. When questioned closely, it appears that mirrors cause great confusion.

<i>“I went to the toilet and then I couldn’t get out and that’s not the first time it’s happened. I don’t know what’s stopping me finding the door. I don’t know It’s as if they’re laughing at me. I think somebody’s making a joke at my expense. That’s what it seems like. (Brian)</i>	Identifier PCA3.19
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Brian says he understands the mirrors are showing reflections of himself but when he is in a room with mirrors on all sides, his confusion and panic increases.

<i>We were catching a plane and I said I’m going into the toilet and ... I couldn’t get out because there was only 40 people, 40 people of me. You must have come across them. So, I started to panic, and I was going up to the door and it wasn’t a door. I was pushing on the glass and I just couldn’t, there didn’t seem to be anyway out. ... I was pushing the wall, you know, glass, mirror, where’s the way out, where’s the way out? I couldn’t find the door. ... I was there, and Barbara was there, and we looked at each other and she said what are you doing? ... and I saw Barbara and I said ‘Help!’ (laughs)” (Brian)</i>	Identifier PCA3.20
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Barbara also recalls the incident.

<p><i>“He got stuck in the toilet in Manchester Airport once. ... He said there were mirrors all the way round and he couldn’t work out which was the door to get out or which was the door into a cubicle or which was a mirror. ... He was quite upset when I found him” (Barbara)</i></p>	<p>Identifier C3.34</p>
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Brian also tends not to use lifts because they often have mirrors in them

<p><i>“Phew, actually the same, and you get two or three or four people in there but you’re the (pauses) well I don’t tend to use them.” (Brian)</i></p>	<p>Identifier PCA3.21</p>
<p><i>“We’ve been in lifts when they have mirrors on both sides and he said he couldn’t cope. He says it like there are 400 of him” (Barbara)</i></p>	<p>Identifier C3.35</p>

4.4.5.7 Memory and insight

Brian’s memory is still good, but he often loses track of time and place. Barbara bought a talking clock and reality orientation board to help but thinks that the problem is due to the fact that they don’t have a normal schedule.

<p><i>“He loses track of what day it is and because we’re doing so many different things, so I have to remind him.” (Barbara)</i></p>	<p>Identifier C3.36</p>
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Comment: Brian suffers badly from perception problems. Although he does not have classic *mirror agnosia* because he recognises his image, too many mirrors where his image is repeated hundreds of times causes disorientation and distress.
Brian’s memory is good, but he experiences time disorientation even with the help of a talking clock and reality orientation board.

4.4.5.8 Environmental Challenges

The physical environment also brings about daily challenges for Brian and Barbara.

4.4.5.8.1 Balance, walking and fear of falling

Brian's visual and perception difficulties mean that walking and swimming can be a problem. Barbara advises that he continually moved over to the left when doing these activities.

<i>"His actual balance is OK but walking he tends to veer to the left a bit. ... You go straight down the swimming pool. He'd be off to the left. Always the left. ... They know him at the swimming pool and I've had to explain to a lot of them and I tend to swim along behind him saying keep right, keep right, keep right." (Barbara)</i>	Identifier C3.37
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Brian describes it as an invisible person pushing him to the left:

<i>"... when he is swimming it's like somebody is pushing and pulling him to the left. He often says he is aware that somebody else is there. He doesn't see them, he just thinks there is somebody behind him pushing him the wrong way. ... He says it's not threatening, it's just a nuisance". (Barbara)</i>	Identifier C3.38
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The couple do a lot of walking. Brian advises he thinks he is walking in a straight line but constantly veering to the left often means that he ends up walking in the road.

<i>"Barbara or anybody else I'm walking with will say 'why do you keep walking in the road?' Apparently, I'm not walking straight. ... I think I am walking straight." (Brian)</i>	Identifier PCA3.22
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Brian can still manage stairs although he is careful. Barbara has installed night lights in the hall and landing to help him.

<i>"He mustn't be rushed but if he goes slowly, he is all right." (Barbara)</i>	Identifier C3.39
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Barbara notes that Brian gets tired very easily and is usually in bed by 9.00 pm. She thinks he needs an afternoon nap, but Brian is not keen as he has never needed to do this in his younger days.

<p><i>“He gets tired very easily. He goes to bed about 9.00 pm and often I think he needs a sleep in the day time. He always thought it lazy to go to bed in the day time and he’s struggling to accept that he has to have a sleep in the day time.” (Barbara)</i></p>	<p>Identifier C3.40</p>
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Comment: Brian and Barbara still have an active life and frequently go walking or swimming although he now tires easily. Barbara states he continually moves towards the left even though he thinks he is walking or swimming straight. This may be a *visual neglect* problem. He can still manage stairs although he goes down slowly suggesting that he is starting to have *depth perception difficulties*.

4.4.5.8.2 Getting Lost

About two years ago, whilst Brian was still working, he started getting lost. Even though the couple have lived in their home for many years, Brian started to get lost in rooms or be unable to find specific rooms. A change in medication and stopping work seems to have ameliorated the situation.

<p><i>“He was off sick. ... He’d be up and down all night, he’d be wandering around the bedroom banging on the walls trying to find the door and he couldn’t find the bathroom. It was just horrendous. It was all through the night. Neither of us was getting any sleep. I think it only lasted probably a couple of months. I think him making the decision to stop work helped a lot in stopping that happening.” (Barbara)</i></p>	<p>Identifier C3.41</p>
<p><i>“For a while he couldn’t find the kitchen and we’ve lived here for years and years and years. But its settled down now. He is on different tablets, so maybe it’s the tablets. I don’t know what helped but it is much better now.” (Barbara)</i></p>	<p>Identifier C3.42</p>

Brian puts the problem down to stress.

"... but it's not as bad as it has been. It's only when I get stressed." (Brian)	Identifier PCA3.23
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Getting lost outside when travelling to client houses had been one of the original indicators showing that Brian was having visual problems. Barbara is aware that Brian will get disorientated in unfamiliar places so now he only ventures out locally with a friend who lives nearby. This person has tAD so them both getting lost together is a slight worry. However, the pair only walk around the neighbourhood and Barbara is comforted by the fact that they are well known in the area and neighbours will help if they get into difficulties.

<i>"He can go to the corner shop over there or maybe a neighbour's house but that's all. He wouldn't be able to find his way if it was somewhere he doesn't know." (Barbara)</i>	Identifier C3.43
<i>"... they are both well-known round here. People know them so if they are found wandering they can be sent in the right direction." (Barbara)</i>	Identifier C3.44

Brian speaks about being frightened of getting lost but thought this problem is improving.

<i>"I have got lost, not so much lately so I'm not going to say I'm getting worse on that, in actual fact, I think I'm getting better. But I do get frightened. (Brian)</i>	Identifier PCA3.24
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Comment: Brian experiences [environmental agnosia](#) and gets lost outside as well as within the house he has lived in for 38 years. Barbara doesn't seem too worried about him getting lost in the neighbourhood as he is well-known, and she feels that neighbours would ensure he gets home safely. This trust enables Brian to keep his freedom to get out and about but also puts him at risk if he wanders too far from home.

4.4.5.8.3 Light and dark

Brian and Barbara spend a lot of time in their conservatory which is bright and airy although, similar problems to the ones they had with mirrors sometimes occur. It has taken time for Brian to get used to the reflections in the windows of the conservatory during the evening. Barbara explains:

<i>“It’s so light in the conservatory, he really likes that, but at night it can be a problem because of reflection. It’s all the way round and he gets really bothered with reflection. He just gets confused. He knows now that there are reflections round there, but he used to sort of say ‘oh there’s a car there’ cause it’s weird if you’re sitting there, if you are sitting at the table, you can see the bin lorry. It looks like it’s going through the garden but it’s going past out here and its reflected there because it’s in the garden behind. So, he’s used to it now. (Barbara)</i>	Identifier C3.45
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4.4.5.8.4 Colour and Contrast

Brian identifies his bathroom products by colour as he is not able to read the labels. The couple both state that contrast is helpful to Brian but hadn’t noticed if any particular colour made a difference for good or bad. During the conversation it is interesting to note that Barbara bought Brian an olive coat with a bright orange liner which helps him put the garment on properly and she also has used an orange bump sticker on the hob to help Brian identify which switch to use. Perhaps his success in these incidences is due to the contrast of colours or the brightness of the orange colour itself. Barbara’s sister, who is an occupational therapist has mentioned that colours might help but the couple are not convinced.

<i>“Contrast helps but there doesn’t seem to be any particular colour that helps. My sister has suggested that and he can’t think of a colour that helps or doesn’t. I got him a coat, nice warm cloth cause we do a lot of walking and it’s a sort of olive kaki colour and it’s got a bright orange lining and fur inside the hood so he always gets that the right way out because of the bright orange lining he can see that easily.” (Barbara)</i>	Identifier C3.46
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Comment: Brian prefers bright natural light but reflections in windows can be disorientating. They have not noticed that any specific colours are useful for Brian’s vision. Contrast seems more helpful – bright orange against a Kaki colour.

4.4.5.8.6 The use of devices to promote independence

Barbara has purchased several products to aid Brian’s independence with various levels of success. These include audiobooks, easy to use mobile telephone, cordless vacuum cleaner, talking clock, bump stickers, a reality orientation board and a contactless debit card.

<i>“He didn’t want to use the reality orientation board to start with. He said they look silly and like he’s got a problem.” (Barbara)</i>	Identifier C3.47
<i>“He’s tried audio books, but he can’t follow the train of it at all.” (Barbara)</i>	Identifier C3.48
<i>“He was getting into a bit of a mess over ...which knob related to which ring on the gas hob, so I’ve put a little orangey red dot on the knob for the ring that he always likes using and I solved that.” (Barbara)</i>	Identifier C3.49
<i>“He hoovers. He gets in a tangle with the cable so that’s why we have got the cordless one.” (Barbara)</i>	Identifier C3.50
<i>“His mobile has two buttons across the top and it’s me and our daughter and he can work that. ... and he can do that. He takes that phone ... everywhere so he can get me.” (Barbara)</i>	Identifier C3.51
<i>“contactless, yes, it made me feel good, very good.” (Brian)</i>	Identifier PCA3.25

Comment: The couple make use of gadgets to help Brian continue to function as independently as possible.

4.4.6 Support and Relationships

4.4.6.1 Family

The couple have one daughter who does not live at home, but she supports them when she can.

<i>“Our daughter lives ... 20 miles away. She says she couldn’t cope with living with him, but she will take him for a walk or have a cup of coffee with him while I go and do something else. So, for a short time, she’s very good with him. (Barbara)</i>	Identifier C3.52
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Barbara’s sister is also very supportive, allowing Barbara time out from caring. Her skills as an occupational therapist who understands the difficulties caused by PCA are proving invaluable.

<i>My sister, the one whose an OT, she’s very good with him. ... we’ve sort of got an arrangement that she comes over once a month ... and stays for a long weekend and I do some voluntary work at the National Trust on Tuesdays so ... she stays and goes for a walk with Brian. They go to the local garden centre or somewhere for lunch and she is very good with him because she understands it completely. (Barbara)</i>	Identifier C3.53
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Unfortunately, other members of the family are not so knowledgeable and are apt to suggest activities which Brian is unable to do.

<i>“His cousin said, “well you should read some newspapers and get some ideas of current affairs, so you’ve got some opinions... I thought, it would be very nice if he could read a newspaper. ... His cousin has no idea at all” (Barbara)</i>	Identifier C3.54
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4.4.6.2 Friends

Unfortunately, Brian has lost contact with most of his old friends and Barbara is aware that he is often lonely.

<i>“... he needs company. He hasn’t got anything he can do on his own” (Barbara)</i>	Identifier C3.55
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They have made friends with a couple in the local neighbourhood. The husband has tAD. Even though there was an age gap, the two wives support each other, and the men have become close friends.

<i>“He’s a really nice guy ... yes he is. Actually, he doesn’t look old. He’s good (Brian)</i>	Identifier PCA3.26
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4.4.6.3 Health and Social Care Professionals

Barbara has found that most of the health and social care professionals they have met are not aware of PCA or the problems it causes Brian.

<i>“Well, our GP at the practice. He didn’t know anything at all about PCA, so I told him about it and he said later that they’d actually had a training session at the surgery so everybody in the Practice knows about PCA now.” (Barbara)</i>	Identifier C3.56
<i>“People with it are telling the experts. It’s just amazing that so many people don’t know about it.” (Barbara)</i>	Identifier C3.57

Barbara discovered a carers group at her local GP surgery but hasn’t found it useful.

<i>“I was going to that for a while, but it was no benefit. They had the solicitor come in to talk about doing power of attorney but he said he was very sorry but he wasn’t a specialist in power of attorney, but he was the only one available on the day and the rest of the group knew more about power of attorney then he did and we were discussing it between us ... and this solicitor person was just sitting there. (laughs). That wasn’t very successful.” (Barbara)</i>	Identifier C3.58
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Comment: Brian and Barbara have a supportive family although not all relatives understand Brian’s specific problems. Their friendship network is small, and they rely mainly on each other. The health and social care

professionals they have been in contact with do not seem to have much understanding about PCA and Barbara has taken it on herself to give them information to raise their awareness.

4.4.7 Changing roles within the relationship

Barbara's life has changed considerably over the last two years. She has retired to look after Brian.

<i>"The earliest I could retire was 55 so I retired. I actually retired because I wanted to get the pension, but it actually fitted very well for when Brian needed a lot more company. (Barbara)</i>	Identifier C3.59
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4.4.7.1 Caregiver Stressors

The couple do not have any professional carers helping them and Barbara sometimes feels trapped because Brian's anxiety means he relies on her both emotional and practically.

She admits that at first, she resented having all these responsibilities.

<i>"Well for about the first year I sort of really resented it I was so annoyed about it, but I think I'm just about getting used to it now. I've not had a carers assessment yet." (Barbara)</i>	Identifier C3.60
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Fortunately, the couple are financially comfortable, and Barbara spends a good deal of time planning outings and holidays for them both.

<i>"I do enjoy it. It's very nice doing all these things and he gets various benefits from it. We've enough money to do all these things and with the discount we get it seems silly to complain really. We've been to Paris twice. We're going on the Queen Mary to New York. We're going on the Flying Scotsman. Lots of nice things whilst we can." (Barbara)</i>	Identifier C3.61
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But sometimes she admits she feels imprisoned by her circumstances.

<p><i>“It drives me mad. I can’t do my own thing at all. He wants me there all the time. ... Every time he gets in a big sort of state saying I don’t love him anymore because I want to go out and do things without him. I just want to do a few things of my own. I used to go to a Tai Chi class and I needed to leave the house at 9 am and I sort of get myself ready to go and then as I was about to leave oh he couldn’t find his sweat shirt or the bathroom light doesn’t work or he can’t turn the kitchen tap on or something, so silly things that wasn’t really a problem but he was having a problem with it. ... it meant I couldn’t get away on time and that meant I couldn’t go to Tai Chi Class and the lady said it doesn’t matter and she understood, it did matter because everybody was in the middle of their session and I would be walking in in a state. So, I stopped doing that.” (Barbara)</i></p>	<p>Identifier C3.62</p>
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Comment: *Barbara describes feelings of resentment, anger and resignation about their situation. Brian does not have support package so leans very heavily on Barbara for social and emotional support. The fact that she has not yet had a carers’ assessments means her needs have not yet been understood.*

4.4.8 Protecting the person with PCA

Barbara is very aware of Brian’s visual and perceptual problems but tries to keep their everyday lives as normal as possible and Brian has household jobs to do which, although they take him a long time, keep him busy.

4.4.9 The emotional impact of PCA

Brian expresses great distress about having PCA. It has stopped him doing most of the activities he enjoys leaving him afraid and sad.

<p><i>“And that’s [PCA] the worst thing that could have happened to me.” (Brian)</i></p>	<p>Identifier PCA3.6</p>
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Barbara is a lot more guarded in showing her emotions but does express feelings of frustration and resentment.

<i>“It drives me mad.” (Barbara)</i>	Identifier C3.63
<i>“I can’t do my own thing at all” (Barbara)</i>	Identifier C3.64
<i>“I had visions of when I retired I would be doing things like a day’s volunteering doing dry stone walls with the National Trust. Things like that. I can’t do any of that.” (Barbara)</i>	Identifier C3.65

Comment: Brian is open about his feelings of distress about having PCA. Barbara uses words such as ‘drives me mad’, ‘can’t do a thing on my own’ suggesting that she is feeling trapped by circumstances.

4.4.10 Facing the future

When I ask about the future, Brian appears resigned.

<i>“I’ll probably just fade away” (Brian)</i>	Identifier PCA3.28
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Barbara is more pragmatic but as his sole caregiver, had some concerns about the future.

<i>“I do wonder what will happen. I don’t know what on earth would happen if I was ill.” (Barbara)</i>	Identifier C3.66
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They have discussed Brian’s future care together. Brian has bad memories of his mother, who had dementia, living in a care home and had made his views clear to Barbara that he does not want this happening to him.

<i>“He said, he’s absolutely (pause) he would kill himself if he was sent to a home. He said he’s going out of here in a box. He’s not going anywhere else. ... I wouldn’t want him to go into a care home unless he was in a state where he doesn’t know me, he doesn’t know where he is or who he is or anything.” (Barbara)</i>	Identifier C3.67
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Barbara’s mum died in her 90s and living in her own home with the help of paid home care. Barbara thinks that this would also be the best option for them.

<p><i>“We’ve thought about it [care home] but sort of ruled it out really. I mean, my plan with mum was that if she did need more care, we’d pay for care. She had a gardener and a cleaner and used dial a ride bus to take her shopping. We just increased the level of care as she needed it, but she stayed in her own house. That’s what we are thinking we’ll do.” (Barbara)</i></p>	<p>Identifier C3.68</p>
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Comment: *The couple haven’t thought about the future to any great extent. Brian is clear that he does not want to go into long term care. Barbara is focusing on how her mother managed at home paying for support such as cleaners, gardeners etc going in when necessary. She believes they will do the same as Brian needs more help. However, her mother did not have dementia and lived very independently to a late age so perhaps Barbara’s views around support will change over time.*

4.4.11 Summary of Brian and Barbara’s story

Brian and Barbara describe several symptoms of PCA they have experienced. This are outlined in [Table 19](#) below.

Table 19: Brian's PCA symptoms

●	Acalculia
●	Agraphia
●	Alexia
●	Apraxia
●	Dressing apraxia
●	Environmental agnosia
●	Hallucinations
●	Simultanagnosia

Brian has had visual and perception problems for six or seven years with a diagnosis of PCA for 18 months. The couple first noticed something was

wrong when Brian's writing skills deteriorated, and his driving became erratic. He started getting lost and perceiving tarmac or patches in the road as solid objects or people.

At first, he was diagnosed as having anxiety. The optician did not notice anything particularly problematic with his eyesight. Brian started to get lost going to client houses and eventually stopped work. His sister in law, an occupational therapist was the first one to mention PCA but at first brain scans were inconclusive, but he finally received a diagnosis of AD with PCA. He was still driving until a potentially serious incident happened when he got disorientated going around a roundabout.

Brian can still undertake his own personal care but needs assistance in recognising washing products using colour to identify them. He has difficulty dressing and finding clothes in cupboards. Brian is still able to eat independently. He undertakes household chores and Barbara has invested in several gadgets to help him remain independent, such as easy to use telephone, cordless vacuum and talking clock.

Brian has great difficulty reading and writing. Unfortunately, audiobooks have not been successful as he finds it difficult to concentrate. He can no longer work out money but has just started using a contactless credit card to buy simple things such as a cup of coffee and this is improving his self-esteem. Barbara was given this tip from a member of the London PCA support group which they had managed to attend.

The couple are still avid walkers and swimmers although Brian tends to veer to the left although he thinks he is going straight. He can still manage stairs, but depth perception problems mean he needs to do this carefully and slowly. When out and about he can get lost and he even has got lost in the house he has lived in for 38 years. He still goes out on his own around the neighbourhood and Barbara believes that neighbours will ensure he will get home safely if he gets disorientated.

Brian prefer bright, natural lighting although reflections in windows can be disorientating. The couple haven't noticed any colour that is helpful in improving his vision, but strong contrasts are helpful.

The couple have a supportive family but rely on each other most of the time. This means that Barbara doesn't get much of an opportunity to have time alone to relax. Barbara has found that most health and social care professionals they have met do not understand PCA and she has provided them with information to raise awareness of the condition.

Brian is open about his feelings of distress of living with PCA. Barbara is more guarded with her emotions but describes feelings of resentment, anger as well as resignation about the situation. She has not yet had a carers assessment, so her needs have not yet been recognised.

The couple haven't thought about the future to any great extent although Brian is clear that he does not want to go into long term care. Barbara remembers how her mother managed at home paying for support such as cleaners, gardeners etc going in when necessary. She believes they will do the same as Brian needs more help. However, her mother did not have dementia and lived very independently to a late age so perhaps Barbara's views will change over time.

4.5 Case Study Four: Laura and David's Story

Because at the time of the interview Laura's PCA was already advanced, she had poor insight and communication skills. David asked if he could be with her during her interview. As the interview progressed it seemed that Laura did not have the ability to understand my questions and often echoed David's responses. This interview lasted for about 30 minutes and was interrupted by visitors who came to sing to Laura. David asked for his interview to take place in a local hotel. This was undertaken in two parts, over two days. Each of these interviews lasted approximately half an hour. The second interview was interrupted by the arrival of David and Laura's social worker. David insisted that the social worker be present during our discussions. Each interview was recorded and fully transcribed.

4.5.1 Setting the Scene

Laura is a friendly, 66-year-old lady who had been diagnosed with PCA for seven years. She had an adventurous streak when younger, driving alone to [place] and back. Laura is well educated, with three degrees: sociology; criminology; and teaching. She started her working in banking. Following university, she became a teacher of children with learning difficulties and then a careers guidance advisor. Laura met **David** in the late 1990s when they were both getting divorced. The two moved in together, finally settling in their present home. Laura's PCA symptoms progressed quickly and her condition now resembled tAD. She appears to remember incidents in her life that have an emotional connection, but these memories are quite jumbled. Laura does not recall taking her degrees or most of her working life. However, the memories of the pupils she taught are still very much in her mind and she speaks about them with affection in the present tense. Laura attends a day centre once a week and her reality is that this place is a school and she is the teacher. Her partner, David is 62 years old and worked as a civil servant before he took early retirement. He suffers from atrophy in his left hand. A year ago, he experienced a stroke but had recovered well. During this time, Laura was admitted into residential care.

David fought for her to be allowed to come back into the community. She has now returned to the family home and David has resumed being her main caregiver. They have professional carers coming in during the week on Wednesday and Thursday to support Laura and give him a break. Unfortunately, David believes that the enforced separation from him has caused Laura to become very agitated whenever he leaves her side and the situation is causing a great deal of tension.

The couple live in a small terraced house situated on a busy road which David is trying to adapt to meet Laura's needs. They live in the downstairs rooms as building work was still in progress upstairs where David is trying to fix up the bedroom to enable carers to stay in the property overnight. The house is very untidy and cramped. The front room has been made into a bedroom with two single beds. The walls are painted in two shades of light blue. Primitive style paintings of bright colours are on the walls in both the bedroom and sitting room. These two rooms are separated by a toughened glass door. The sitting room is filled with easy chairs, books and a large TV and there isn't much room to move around easily. There is a small table next to Laura's armchair. Lighting, both natural and electric is poor in both the bedroom and sitting room. A small extension has been built onto the back of the property which contains the kitchen and accessible bathroom with shower. The lack of space in the home means that the use of a walking frame or wheelchair is impossible.

4.5.2 Realising something was wrong

David explains that in 2007, they both realised something was wrong when Laura was trying to address Christmas cards. Laura found she couldn't write properly. Then she started having difficulties in using an ATM machine.

<p><i>“... first realisation was when she was trying to address Christmas cards. She found she couldn't write properly. ... I remember one time we were in [name of place] r, and she managed somehow or other in a cash machine to put her card into the bit where the receipt comes out. God knows how she did it, but it went into the machine. We laughed about that.” (David)</i></p>	<p>Identifier C4.1</p>
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They had both laughed at the incident at the time but then Laura started falling over:

<i>“She kept tripping over her legs” (David)</i>	Identifier C4.2
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The situation continued to get worse at Laura’s workplace.

<i>“all the work was on computer keyboards, and this is when the deficiency came out. She couldn’t hit the keys. She was slow, slow. She couldn’t take the phone calls and write records, and this is how we noticed her inefficiency.” (David)</i>	Identifier C4.3
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Laura’s employers started proceedings to sack her for inefficiency.

<i>“It was so sad. We didn’t know what the problem was, we just knew there was a problem when she couldn’t use the phone anymore.” (David)</i>	Identifier C4.4
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Laura was still driving. Then one day when they were driving home, she began to panic.

<i>“it was a bendy road and she got fazed. ... She said, ‘I can’t do this anymore’ and that was about the last time she drove. It was the bends. Spatial awareness. We didn’t know then, but we know now. (David)</i>	Identifier C4.5
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Comment: Laura’s is displaying poor motor skills making it difficult to use a computer and telephone and ‘tripping over her legs’ which may be due to [apraxia](#). Visual perception problems may also be causing problems with writing ([agraphia](#)) and using the phone. It is interesting she still drove for some time even with all these visual and motor skills difficulties.

4.5.3 Journey to diagnosis

They went to see their GP and after some tests, Laura was sent to a senior specialist in AD. They saw two different consultants. They went twice for tests and at the third visit, in 2010, they were shown a model of a brain and Laura was given her diagnosis.

<i>“her memory was good then” (David)</i>	Identifier C4.6
<i>This doctor ... he did these tests with her. And he’s the one who gave us the opportunity to go to [name of place]. ... he was a senior specialist in Alzheimer’s’. We saw two different consultants. I think we went twice and the third time we went ... they diagnosed her with Alzheimer’s and I was not surprised. ..., and they said you’ve got PCA. We said, what’s that? (David)</i>	Identifier: C4.7

Comment: This diagnosis seems to have happened quite quickly possibly due to the specialist experience of the neurologist.

4.5.4 Reactions to knowing

The couple had never heard of the condition and the specialist explained the difference between tAD and PCA.

<i>“They said with most people with Alzheimer’s, it’s the front of the brain which is affected. You’ve got this sort with spatial awareness and vision problems and that’s at the back of the brain and this is what this model was all about. Different colours. ... I was not surprised” (David)</i>	Identifier C4.8
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David immediately phoned her employers and the industrial tribunal was stopped with Laura being retired on medical grounds at the age of 60 years.

<i>“The same day, my boss said, there’s a package going if you want it and that was it, I never went back to work.” (David)</i>	Identifier C4.9
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Laura also visited a local optician who sent her to the eye hospital to be registered visually impaired. They did not find this an easy process.

<i>“We went to see him a couple of years ago because I was trying to get her DLA upgraded and that was an experience. ... I kept saying the forms don’t fit Alzheimer’s basically. To put it bluntly if you’ve got four limbs as far as DWP is concerned you’re a fully functioning person unless you fall into the category of vision and she didn’t fall into that category the first time. But I thought I’m not going to give up on this and I kept saying look mobility, tripping over kerbs, crossing roads. In fact, the guy from the job centre came up to the house and filled the form in for me and we still got knocked back. ... the last time we went to the consultant I said, ‘can you do me a letter explaining what PCA means so I can give it to the DWP. He wrote a far more detailed one to the GP which I’ve got as well so I sent that off to the DWP and then we got the visual impairment thing signed off.” (David)</i>	Identifier C4.10
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The consultant told them to go out and enjoy themselves whilst they could. *“make the best of your time”*. Over the next few years they went on Safari in Kenya, even though Laura’s mobility was starting to slow down. They also enjoyed two cruises. David sighs:

<i>“we’ve done as much as we can.”</i>	Identifier C4.11
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They continued to visit the specialist for annual check-ups until this last year when the journey became too difficult for Laura and they now see their local consultant.

Comment: *The difficulty in getting Laura’s DLA upgraded appears to be due to the Department of Work and Pensions (DWP) not fully understanding how PCA affects individuals.*

4.5.5 Day to Day challenges

Talking with Laura, she advises me she is perfectly OK and can do everything. However, PCA is now causing global incapacities.

4.5.5.1 Personal Care

David advises that he now must support Laura with all her personal care. The professional carers help a couple of days a week but otherwise David undertakes all these tasks. Laura believes she can still do this for herself and becomes very agitated when trying to express her needs. This causes David a great deal of distress.

<p><i>“I’m quite able to go along and do it on my own.” (Laura)</i></p>	<p>Identifier PCA4.1</p>
<p><i>“I try and bathe her on the occasions when it’s down to me, cause she was terrible. She used to be awful and have a go at me and that was the most stressful part of the day. I’ve turned it into a game now and I always start off by putting the shampoo on her hair to get her wet from the beginning.” (David)</i></p>	<p>Identifier C4.12</p>
<p><i>“She can’t recognise shampoo. I know she said she could. What I do is put the shampoo on her and I sort of say do you want to help me by doing your arms and things like that. She can do that sort of stuff if I prompt her but that’s the limit. Getting her dry. She always thinks she’s wet and that can be difficult.” (David)</i></p>	<p>Identifier C4.13</p>

Comment: Laura’s condition has progressed into many of the symptoms of tAD including challenging behaviours. Her resistance to David supporting her to shower could be because, as she states she can ‘do it on my own’ which is her reality. Not recognising the washing products may be due to reading difficulties (*alexia*) or *associative agnosia*.

4.5.5.2 Dressing

Laura has not been able to dress herself for two or three years.

<i>“She can actually help me to put pullovers over her head and things like that and by mistake I got some ... trousers recently that had a zip in them and a button in and she actually managed to get the button undone which I thought was an achievement.” (David)</i>	Identifier C4.14
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Comment: David is describing [dressing apraxia](#).

4.5.5.3 Food Preparation

Laura says she quite likes cooking. Interestingly, when asked if she ever has any difficulties in doing these tasks, she starts to talk about problems her students face.

<i>“But some of the students might need some help in terms of what to do, what they are going to do.” (Laura)</i>	Identifier PCA4.2
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David remembers Laura’s cooking and baking but advises it is a long time since she has done anything in the kitchen, but he has kept all her cookery books.

<i>“You saw the cookery books. ... I’ve deliberately kept things that I’ve written down. I think they’re good memories for me. So, there’s recipes and stuff like that.” (David)</i>	Identifier C4.15
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Comment: *Laura’s physical and visual incapacities now make it impossible for her to undertake cooking or baking. Laura phrases her words in an interesting manner. I had asked her if she needed help to cook and bake but she talks about her students needing help. Is she actually talking about herself and identifying herself needing help but within the context of her students?*

4.5.5.4 Eating and drinking

Laura can no longer use cutlery and David must support her to eat.

<i>“She can’t eat properly. I have to feed her. She can eat finger food but anything else ... It’s been coming for a long time... we’ve given up really.” (David)</i>	Identifier C4.16
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He has noticed that she does not see very well on the right-hand side and, although Laura is right handed, he is trying to encourage her to use her left hand.

<i>“... I’m trying to get her to use her left hand. She seems better off with it actually and there’s this thing about her right-hand vision.” (David)</i>	Identifier C4.17
<i>“we know her vision on the right-hand side is the worst. It’s a weak point. She misses things on the right side, so I always try to give her things to the left. She has trouble finding things on her plate.” (David)</i>	Identifier C4.18

Comment: Laura appears to have right sided *visual neglect* or missing things on the plate could be due to not being able to process a full visual scene properly (*simultanagnosia*). She may be seeing separate elements on the plate but not able to put them together as a whole picture.

4.5.5.5 Reading and numeracy

Laura can no longer read or write or understand numbers or money.

<i>“She doesn’t read. I always used to get her Bella and Best. She used to like reading them and the supplements from the Sunday Papers’, but she’s stopped doing that years ago. I’ve thought about talking books. I’ve asked her about that before. We’ve never tried it” (David)</i>	Identifier C4.19
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David advises he is always reminding Laura about her three degrees, but Laura does not seem to remember, but she does recall that she enjoys music and that she has been singing with her carers that morning.

A qualitative study exploring the specific everyday social and environmental challenges faced by community dwelling people living with posterior cortical atrophy (PCA)

<i>“[name] University that’s where you got your first degree” (David)</i>	Identifier C4.20
<i>“Was it?” (Laura)</i>	Identifier PCA4.3

<i>“What have you been doing today?” (researcher) “Oh, what have we been doing? Oh, yes, we’ve been having a focus on music. I like singing.” (Laura)</i>	Identifier PCA4.4
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Comment: David describes symptoms of [alexia](#), [agraphia](#) and [acalculia](#). Laura’s memory is also now compromised. Did she remember singing because that causes a happy emotional memory which she can remember?

4.5.5.6 Problems with visual perception

David feels that Laura has trouble perceiving what she is seeing and that if she puts something down she invariably cannot find it again.

<i>“She picks things up and holds on to them more than anything else. I have to watch her with bracelets and things like that. Just in case she puts them down somewhere and can’t see where they’ve gone.” (David)</i>	Identifier C4.21
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Laura spends a lot of time watching TV and she seems to enjoy it. However, when I asked her about TV, she again starts referring to her students and how they need time to understand things. David is not convinced she is able to see the screen clearly.

<i>“... natural programmes are good. ... and they can be (pauses) you think about that in terms of what you’re wanting to do, you know. But some of the students, some of them just need a little bit more time to get things sorted out so that it makes sense and relates to something.” (Laura)</i>	Identifier PCA4.5
<i>“... It’s interesting when she’s watching TV. I’m still not convinced that she actually sees the images properly, if at all.” (David)</i>	Identifier C4.22

Comment: David seems to be describing *simultanagnosia*. Laura again uses the metaphor of her students needing time to make sense of something when referring to herself.

4.5.5.7 Memory, communication and insight

Laura’s long and short-term memory are poor. She appears to only remember incidents that have an emotional attachment to them. She remembers seeing one of the Beatles:

<p><i>“It was at wasn’t it when you saw George Harrison walking his dog.” (David)</i> <i>“Yes.” (laughs) (Laura)</i></p>	<p>Identifier PCA4.6</p>
<p><i>“You drove all the way to [name of place]” David</i> <i>“Yes. (laughs). Well I just decided I would drive there and get on with it, yes”. (Laura)</i></p>	<p>Identifier PCA4.7</p>

But she cannot remember being at University or the different jobs she has done.

<p><i>“And you’ve got two degrees and then you decided to go on and do another one.” (David)</i> <i>“Did I?” (Laura)</i> <i>“Yes. You went and got your teaching degree.” (David)</i> <i>“Oh (laughs) oh! “(Laura)</i> <i>“Oh goodness me. I haven’t done all these things have I?” (Laura)</i> <i>“what did I do, where did I work?” (Laura)</i></p>	<p>Identifier PCA4.8</p>
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4.5.5.8 Environmental Challenges

4.5.5.8.1 Balance, walking and fear of falling

Laura’s energy levels are low, and David explains that she often dozes in her chair. Getting her to exercise is proving difficult and David is trying to think of ways to encourage her.

<p><i>“She doesn’t exercise. ... A physio came round to see us and she turned it into a game cause we’ve put rails in the bathroom and she showed us some exercises but when she went into care everything stopped. I’ve been thinking about getting her back again cause you see the ballet school girls on the barre well that I was thinking as a game. We could put some music on and exercises.” (David)</i></p>	<p>Identifier C4.23</p>
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Laura has problems walking and is very slow at undertaking this activity. Her perception of the road or pavement surface is faulty which is quite dangerous if they are crossing the busy road outside their house.

<p><i>“ ... she just can’t walk very fast. Well she can’t really see either and if there’s a change in the colour of the tarmac or the pavement, she’ll think it’s a hole or something like that. So, she actually spots things that aren’t a problem.” (David)</i></p>	<p>Identifier C4.24</p>
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She is also becoming resistant to holding David’s hand when crossing the road which makes her more at risk of falling.

<p><i>“She doesn’t like me holding her hand. That’s only something that’s come out in the last couple of months so if I’m holding her hand to get her across the road or something, she keeps saying that’s mine, let go, and it’s becoming a bit of a problem. She keeps her hand in her pocket and I’m trying to link on it. Last night, we were going across the road, it takes us so long to get across the road where we are with traffic coming. Something happened. She didn’t slip, but it shook me, and we were still on the road. I had to make sure we were alright. But it’s that sort of thing that’s a bit worrying. I have got a white stick that the ROVI gave us and I’m thinking of taking it out myself but some of the drivers round here. They wouldn’t bother anyway.” (David)</i></p>	<p>Identifier C4.25</p>
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Comment: Laura’s lack of exercise will be having a detrimental on her overall health and cause muscle weakness which in turn will diminish her ability to walk. Her behaviour in refusing help from David could be because she thinks she can walk without assistance and is trying to assert her independence.

4.5.5.8.2 Getting lost

Because of Laura’s mobility problems, she is at a very reduced risk of wandering off or getting lost. However, it is interesting to hear Laura’s response when I ask her if she ever got lost. It appears even though she cannot move around by herself, she sometimes feels disorientated or lost within herself.

<i>“You know, you are not quite sure where you are going to, but you need some help to find out where you can go” (Laura)</i>	Identifier PCA4.9
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4.5.5.8.3 Light and dark

The couple’s house is quite dark. There is a lack of natural light, particularly in the middle sitting room and the electric lighting is not bright. David explains that they have put in the toughened glass door between the bedroom and the sitting room to help the situation, but the area appears to be quite hazardous.

<i>“The rest of the place I’ve tried to make it as bright as possible. We’ve a problem with the middle room. That glass in the doors from the bedroom to the living room into the kitchen. That glass is safety glass, toughened. I know because I slipped on a bloody coat hanger one night and I went crash against the glass. I got a bad cut on my head, but it didn’t go through, so I know it’s safe. It’s slightly tinted for a bit of privacy. The carers say we need more light in the middle room. The electrics are all the in wrong place, so we have a lot of extension leads. (David)</i>	Identifier C4.26
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4.5.5.8.4 Colour and contrast

When conducting David’s interview in the hotel, he points out that that Laura has difficulty walking on the carpet which was full of whirly colours.

<i>“She doesn’t like this patterned carpet. She hasn’t said it, but I can see when we go out this bit here is difficult for her.” (David)</i>	Identifier C4.27
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Within the home David has tried to improve the décor, changing a patterned carpet for a plain one and painting the bedroom walls two tones of light blue.

“We’ve done all this over the last couple of years and like the colours in the bedroom, the blues, that was a bit of fun, That’s sea and sky.” That’s the way that was done. I mean I’ve tried my best with the colours in the house, especially the carpets. they were whirly ones before (David)	Identifier C4.28
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***Comment:** David and Laura’s home is dark and extremely cluttered. David is trying his best to adapt the home to Laura’s visual needs but at present she is at risk of tripping over extension wires that cannot be seen properly because of the poor light levels.*

4.5.5.8.5 The use of devices to promote independence

David and Laura had visited RNIB several years previously and bought a talking watch and large button telephone, but Laura has not found them helpful. They have not tried anything else although David says he encourages people to think about RNIB and their products because they might help other people with PCA.

4.5.6 Support Networks and their understanding of PCA

4.5.6.1 Family

It appears their family members are not particularly supportive regarding Laura’s condition. To start with her late father did not believe there was anything wrong with her.

“He said, ‘she’s normal’.” (David)	Identifier C4.29
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Laura’s brother only visits occasionally and thinks she will be better off in a care home. David has finally persuaded him that, in the short term, Laura should be looked after and have a better quality of life, at home.

<i>“Her brother has done absolutely nothing. The only thing was when the inheritance [Laura’s parents] came through, he wanted to know what I was spending it on. I said however it’s spent it’s all going to be spent to make sure Laura has the best possible time.” (David)</i>	Identifier C4.30
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4.5.6.2 Friends

David advises he has one friend who supports him, and he pays for people to come in to entertain Laura with singing each week. Apart from this, the couple seem quite isolated.

David thinks it would be helpful if they could be part of a PCA support group. They live too far away from London to attend that group and he would like to organise one nearer their home but there are only a few people he knows of who were dealing with PCA in the North, that could possibly attend.

<i>“We don’t know anybody else, so I thought about, you know they’ve got these splinter groups, support groups down South and it’s not feasible to go down there but maybe you can have a Northern one covering North East and North west and we could meet up halfway. That kind of thing.” (David)</i>	Identifier C4.31
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4.5.6.3 Health and Social Care Professionals

Laura attends a day centre which is very helpful regarding her condition. It is a mixed group of people, younger people with learning difficulties (to whom Laura relates) and those with dementia. Laura is the only person there who has visual difficulties.

<i>“She goes to Red house There’s a guy there who has Downs and he relates to Laura. She likes going there. She regards them as her students and it’s a teaching day. So, she feels she’s giving something.” (David)</i>	Identifier C4.32
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To help Laura, the health and social care professionals the couple liaised with, including the local optician, have been made aware of the nature of PCA by David as initially they had not heard of the condition.

<i>“I recently sent ... a package of information to Specsavers on PCA which they’d never heard of so that would be discussed at their Staff Meeting.” (David)</i>	<i>Identifier C4.33</i>
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<i>“I’ve had to explain to Social Workers what it is.” (David)</i>	<i>Identifier C4.34</i>
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<i>“The optician ... was fascinated. Well she didn’t know what to do and she referred us to the eye specialist ... (David)</i>	<i>Identifier C4.35</i>
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David has been in touch with a local journalist and he is hoping they will run a story about Laura and PCA to raise awareness.

<i>“... we do have a journalist interested. She rang me up on Sunday morning and we were on the phone for almost an hour and I was explaining what PCA was to her and she was absolutely fascinated. So, it’s been left that she will do a follow up.” (David)</i>	<i>Identifier C4.36</i>
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Comment: *David and Laura do not have a supportive family and they have few friends. This puts a lot of strain on David to support and care for Laura physically, socially and emotionally. They seem to have a good relationship with their social care team and Laura is accessing appropriate day care and support workers. However, David has found the care system difficult to navigate. He is trying to increase awareness of PCA himself.*

4.5.7 Changing roles within the relationship

Even though David is still recovering from his stroke, he manages to care for Laura undertaking her personal care as well as looking after the home, though he is sometimes in physical distress.

<i>“Like today before I picked you up I dropped off a load of laundry. I do the easy stuff but ironing and that is a bit beyond me. But I’m Ok. The stroke was on the right side and the right side is still better than the left. I have a constant pain in this hand.” (David)</i>	<i>Identifier C4.37</i>
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4.5.7.1 Caregiver Stressors

David says he gets stressed very easily if Laura becomes agitated or upset. However, a key stressor is all the legal problems they have because they are not married. He explains they have both been in bad marriages before and agreed not to marry again. Now, due to Laura’s condition, even if they wanted to marry, they are not allowed.

<i>“we have all sorts of legal problems now because we never got married. We’ve both been married before and we said we wouldn’t do that again and now we’re not allowed to now” (David)</i>	<i>Identifier C4.38</i>
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This leaves David in a precarious position where eventually he might lose the family home if Laura goes into full time care.

<i>“Even when she was diagnosed I kept thinking oh we’ve got to do wills. The only thing we did was Power of Attorney, so we never did our wills, never got married and now they say we can’t do that so I’m not on the deeds of the house either so it’s a total shamble really. These are all sorts of things I did think about, but I didn’t want to push them. That’s why I got the other Power of Attorney [brother and his friend’s partner] and now she has quit the role cause I didn’t want anyone to take advantage and I put clauses saying I could do everything apart from sell the house. I thought that was sensible. It showed I was being honest. That’s what came back to bite us.” (David)</i>	<i>Identifier C4.39</i>
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David advises he is going to go and talk to the GP about Laura's tablets to see if there is anything that might improve her mood but feels her distress comes from fear that he is going away from her again. This situation is stopping him having breaks from Laura which is building up yet more stress within him and not helping his recovery from the stroke.

<p><i>“... it’s been a bad few weeks ... I am going to talk to the GP about the other tablets but a lot of it now seems to be geared to when I go out. Like this morning, she was already getting worked up. ... Obviously, I get upset because I don’t go out unless there is a real reason for going out ... like when it’s my supposed break on Wednesday and Thursdays. But I’ve always said if anything happens in the house I’m always contactable. Like last week when I was in (place) I phoned to let the carer know I was there so if anything happens I just get a taxi straight back ... cause I won’t leave the carers in a position cause the stuff over Christmas was terrible and one of the carers, she hit and (name of carer) is strong enough to say well it happens. It is difficult because I know I’m going out, but I know the people I am leaving her with. So, it’s upsetting but I can’t stay in there all the time. That’s the problem. ... That’s what started to happen last year. I get upset. ... I get very very stressed. Well it’s better if I can go out but I can’t go out. I just try to sit down and just ignore it. Let my levels get back to normal.” (David)</i></p>	<p>Identifier C4.40</p>
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Comment: *Because of Laura’s increasing challenging behaviours, David’s stress levels are high, and this is detrimental to his own health issues. The problems of not being married highlights the benefits of timely diagnosis to allow time for couples to plan ahead and get advice regarding their legal rights for long term care provision and end of life scenarios.*

4.5.8 Wanting to protect the person with PCA

Showing me his hand, David muses:”

<i>“she looked after me then and it’s my turn now. But I always tell her that we are getting better together.” (David)</i>	Identifier C4.41
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Before Laura was able to come back home, David had to attend a “best interests” meeting. His former role as a civil servant has helped him manage all the bureaucracy.

<i>“I’m just doing it from the other side of the table. So, everything I’ve applied for I’ve got, including a parking space outside the house which a neighbour doesn’t like but it’s there now.” (David)</i>	Identifier C4.42
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4.5.9 The emotional impact of PCA

The emotional strain on David is noticeable and his social worker voices that they do worry that too much stress may result in him becoming ill again. David explains that sometimes Laura’s remarks to him are very hurtful. David believes this behaviour appears to stem from the time she was in care.

<i>“Even though she says she hates me and she wants me to die and all the rest of it, there is absolutely no doubt she is totally dependent on me being around and I think that’s what it is all about. This stuff is coming out now because she thinks I am not going to come back.” (David)</i>	Identifier C4.43
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During our interview, Laura seems quite happy but rather distant although at one point she turns to David for reassurance.

<i>“Do you think I’m going to be alright?” (Laura)</i>	Identifier PCA4.10
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To which David responds:

<i>“we are getting better together.”</i>	Identifier C4.44
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Comment: David gives the impression of being very upbeat most of the time but admits to becoming increasingly stressed by Laura’s behaviour and comments. Laura’s move from home to long term care to home again would have been emotionally stressful for her and her agitation could be due to fear and uncertainty at a deep level even if she doesn’t express it when we talk.

4.5.10 Facing the future

Laura does not answer when I ask her about the future but at my meeting with David, he speaks about being realistic and knowing that Laura would one day go back into care. He also says he has taken precautions, but would not elaborate, that she will be looked after if anything happens to him.

<i>“Her brother had given her a year, two years ago before she was taken into care or had to go into a home and despite what he said I’ve always been realistic about this that one day she will have to go into a home. ... She really hated it in those homes, you could see she was upset. But the next time, unless something dreadful happens to me and we’ve done a contingency plan for that. Unless something happens to me like that, but we’ll cross that bridge if it did happen, but I said to her brother look give her a chance to enjoy what she’s got whilst she can because when it comes to the point when she has to go into a home she won’t know. ... and he accepted that and that’s the way that I got him on side to let Laura come home.” (David)</i>	Identifier C4.45
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Comment: *David seems realistic about the future and appears to have planned ahead in case he dies before Laura although he appears not to have put much consideration into his own future social and care needs.*

4.5.11 Summary of Laura and David's story.

Laura presents with several symptoms of PCA as noted in [Table 20](#) below:

Table 20: Laura's PCA symptoms

●	Acalculia
●	Agraphia
●	Alexia
●	Apraxia
●	associative agnosia
●	dressing apraxia
●	Simultanagnosia
●	Visual neglect

She is also experiencing tAD symptoms including word finding difficulties, delusions and behaviours that challenge.

Laura's visual/perceptual problems started with difficulties using a computer, telephone and writing. She also displayed poor motor skills including tripping up. Even though she was displaying all these worrying symptoms she continued to drive for some time.

Even though her memory was good at this time she was quickly referred to a specialist who was knowledgeable about PCA. She was diagnosed with the condition which was explained well to the couple.

Laura's condition now displays many of the symptoms of tAD including loss of insight, poor memory and behaviours that challenge. She can also no longer read, write or understand finances, dress herself or independently eat. David needs to support her with all her personal care needs.

Although the couple have good support from their local social services, they found it difficult to claim for upgraded DLA as the DWP were not aware of the difficulties experienced by PCA.

Laura has a poor memory for facts but seems to be able to recall incidents that have an emotional connection and she still enjoys music and singing. She often refers to her students as needing help to do or understand things rather than herself needing the support.

Laura's poor walking ability may be being exacerbated by her sedentary lifestyle. However, she insists she can walk independently often refusing help from David which increases her falls risk. Their home has low light levels and is extremely cluttered making Laura at extra risk of falling.

The couple have poor family relationships and few friends which puts a lot of pressure on David to care for Laura's emotional, social and physical needs whilst still recovering himself from a mild stroke. Laura does have a care package and attends a day centre once a week and has support workers two days per week.

Because the couple are not married, David is in danger of losing the family home if Laura goes into long term care. It appears David has a plan in place so that Laura is looked after if he becomes ill or dies but he does not seem to have considered his own future needs. Because of Laura's increasing challenging behaviours, David's stress levels remain high and long-term this will be detrimental to his own health. The problems of not being married highlights the need for early diagnosis to allow time for couples to plan ahead and get advice regarding their legal rights for long term care provision and end of life scenarios.

4.6 Summary overview of all interviews

The four interviewees with PCA have a diagnosis of between 18 months and 7 years and are at different stages of the condition. Polly and Joe are not clear how long Polly’s PCA symptoms have been occurring because she has had poor sight since she was young and when she was referred to the eye hospital it wasn’t determined for some time whether her initial vision problems were due to glaucoma rather than PCA. **Table 21** shows that the interviewees have experienced deteriorating vision for several years before finally being diagnosed with PCA.

Table 21: Interviewees with PCA

Name	Onset of Symptoms	Age at onset of symptoms	Year of Diagnosis	Time between onset and diagnosis	Age at diagnosis	Timeframe of diagnosis of PCA to time of interview
Betty	2004	61 years	2015	11 years	72 years	2 years
Polly	1970 (?)	25 years (?)	2011	21 years (?)	67 years	6 years
Brian	2010	54 years	2015	5 years	59 years	18 months
Laura	2007	57 years	2010	3 years	60 years	7 years

Using the stages of PCA model (Crutch, Walton, Carton, & Shakespeare, 2017), **Table 22** shows that all four individuals have symptoms consistent with Stages 2, 3 and 4 and varying aspects of Stage 5 of the disease. In addition, Laura appears to have moved onto Stage 6 and was displaying symptoms of tAD.

The next section will amalgamate the findings from the four case studies to explore the overall everyday challenges of living with PCA. By examining the similarities or differences in the lives of these people, it is envisaged that a better understanding of the impact of this condition as practical and emotional perspectives start to emerge.

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Table 22: Presenting symptoms of interviewees with PCA

Symptoms of PCA*	Betty	Polly	Brian	Laura
Stage 2				
a loss of confidence in their driving ability	✓	✓	✓	✓
problems using a computer	✓	✓	✓	✓
difficulties with reading, writing, spelling and/or arithmetic	✓	✓	✓	✓
difficulty recalling and writing down telephone numbers	✓	✓	✓	✓
difficulty assembling novel devices/objects	✓	✓	✓	✓
The person may experience vertigo or other balance problems	✓	✓	✓	✓
may experience difficulties finding the right words when speaking or may experience odd visual sensations (e.g. occasional changes or washes of colour in the centre or periphery of their vision)	✓	✓	✓	✓
Stage 3				
Difficulty seeing things or only seeing part of something	✓	✓	✓	✓
General 'clumsiness'	✓	✓	✓	✓
Difficulty finding things in a handbag, cupboard, etc. –	✓	✓	✓	✓
Difficulties dressing and performing other activities of daily living that require spatial judgement	✓	✓	✓	✓
Inability to tell the time from a clock or watch	✓	✓	✓	✓
Reading and writing difficulties	✓	✓	✓	✓
Mild memory problems for recent events	✓	✓	✓	✓
Mild difficulty retrieving the correct word in conversation	✓	✓	✓	✓
Stage 4				
Inability to read labels on food packets –	✓	✓	✓	✓
Problems discriminating coins –	✓	✓	✓	✓
Inability to tell the time from an analogue or even digital watch	✓	✓	✓	✓
Dissociation in ability to walk, sit down and stand up				✓
Being able to see some things but not others	✓	✓	✓	✓
Cannot read but can still follow or at least get pleasure from TV or the cinema	✓	✓	✓	✓
Getting lost within the home.	✓	✓	✓	✓
An inability to identify even very familiar faces				✓
Resorting to eating with one's hands, or being fed	✓	✓		✓
increasing word finding difficulties	✓		✓	✓
Stage 5				
Require help with most or all everyday tasks, including toileting and personal care	✓	✓	✓	✓
Experience spatial problems not limited to vision	✓	✓	✓	✓
Require support and/or supervision when walking	✓	✓	✓	✓
Episodic memory and word finding difficulties may also now be prominent			✓	✓
Some people may have problems with multiple senses				
Increased sensitivity to pain		✓		
become more sensitive to the cold	✓	✓	✓	✓
Sense of hearing may change				
Feelings of imbalance or instability	✓	✓	✓	✓
Stage 6				
Experience changes in sleep patterns			✓	✓
Have increasingly frequent trouble controlling their bladder or bowels –				✓
Become quiet and withdrawn				✓
Experience behaviour and personality changes		✓		✓
Have difficulty communicating				✓
Experience continued deterioration of sensory functions				✓
Stage 7				
Loss of ability to respond to their environment				
Unable to carry on a conversation				✓
Unable to control movement.				
Total functional dependence				
Unable to walk				

*as described by (Crutch, Walton, Carton, & Shakespeare, 2017)

5.0 Chapter Five: Findings: Themes

5.1 Introduction

This chapter discusses the key themes that emerged based on the narratives of the study interviews. The outcomes from these theme-based discussions will be compared with the study literature in [Chapter Six](#).

The analysis for the study interviews was mapped into seven themes to highlight the main experiences the interviewees described that were making the most important impact to their lives as they progress through the journey of PCA. These themes are outline in [Table 23](#):

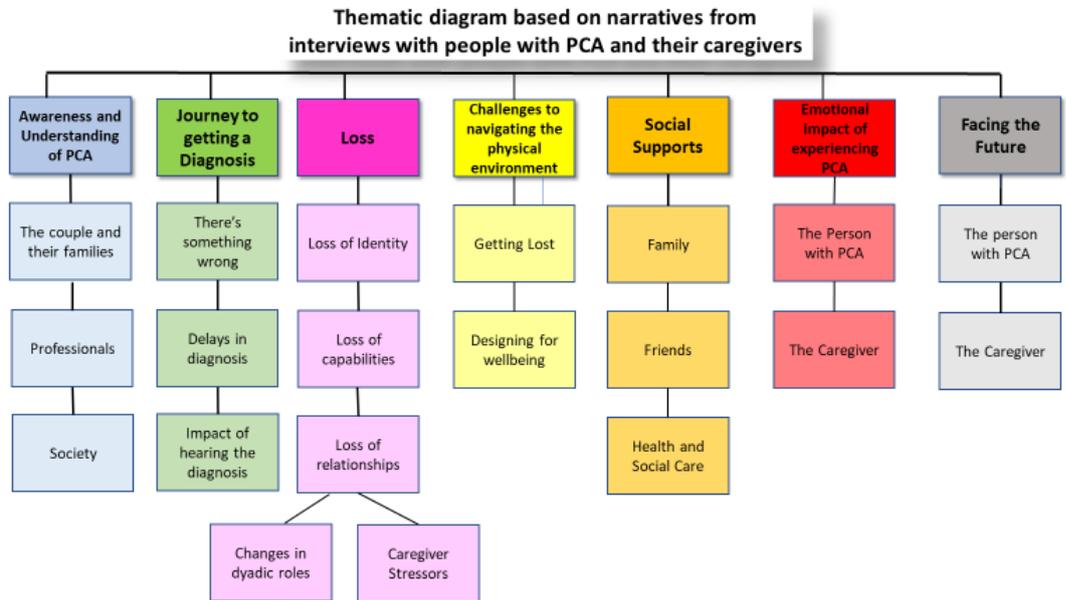
Table 23: Emerging Themes

Themes	
1.	Awareness and Understanding of PCA
2.	Journey to Diagnosis
3.	Loss
4.	Challenges to coping with the physical environment
5.	Social Supports
6.	Facing the Future
7.	The emotional impact of living with PCA

[Figure 3](#) shows how each theme was then divided into sub-themes to ensure all data regarding each particular area of interest could be fully considered.

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Figure 2: Thematic diagram based on narratives from interviews



A larger picture of this diagram is shown in [Appendix Eight](#).

The identifiers referred to in the discussion below refer to the identifiers in the quotes in Chapter Four. The letters PCA or C refer to either a person with PCA (PCA) or a caregiver (C). The first number gives the case study number and the second number the statement. For example:

<i>“I was alright all my working life it was quite normal. It was only just little by little things were going awry (Betty)</i>	Identifier: PCA1.1
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[Identifier PCA1.1](#): The statement was made by a person with PCA (PCA) in Case Study One (1) and it was the first statement made by that person (1) in the text.

5.2 Theme One: Awareness and understanding of PCA

All four cases demonstrate that awareness and understanding of PCA is globally lacking. For the couples themselves, none knew of PCA before diagnosis (PCA.1.7; PCA2.7; PCA2.6; PCA3.3; C4.4).

Because family members do not understand the condition, sometimes it is not believed that there is anything wrong or the couple are given inadequate support (C1.30; PCA2.35; C3.54; C4.29).

The health and social care professionals the couples encounter also appear uninformed. This results in a lack of appropriate advice and lack of assistance (C1.31; C1.34; C2.3; C2.45; C3.56; C4.33).

Some of the caregivers have taken it upon themselves to try and educate others (C3.56; C3.57; C4.33; C4.34). One individual has even contacted a journalist who is going to run a story about the condition in the local newspaper (C4.36).

5.3 Theme Two: Journey to getting a diagnosis

5.3.1 There's something wrong

In all cases, couples state they had noticed something was not quite right either with their own or their partner's vision, long before they began to seek help. Often it was little things that seemed trivial at the time (PCA1.1; C1.1; PCA2.1; C.2.1; C3.1; C4.1; C4.2). This is in keeping with tAD.

Gradually as visual and perceptual capabilities deteriorated, interviewees start talking about the specific problems that were beginning to become noticeable. For example, writing legibly became difficult (C3.28; C.4.1). Of note are their comments about driving becoming erratic. The case study participants are either of working age, or have just retired, as their PCA started to become noticeable. Even so all still drive on a regular basis even though they realise there is something wrong with their vision (PCA1.4; C2.2; C3.2; C4.5)

In some cases, individuals say they stopped themselves driving (PCA1.3; PCA2.3; C4.5) although in one case it takes a serious incident before one participant is forced to stop (PCA3.2; PCA3.5).

5.3.2 Delays in diagnosis

In this study, it appears that diagnosis has not been straightforward in any of the cases. When individuals visit the optician, they are prescribed new

spectacles. Visual and perception challenges that might be due to cognitive causes are not considered by the optician at this stage (C3.4).

The couples next visit their GP. Because their memory is good at that point, it appears that dementia is not suspected in the first instance (C3.5; C4.6), symptoms being put down to stress (C1.4) and medication for anxiety is prescribed.

As vision and perception further deteriorates, the couples are sent to see numerous consultants and specialists (PCA1.6; PCA1.7; C2.4; C3.8) until they finally receive a diagnosis. A delay in the process sometimes appear to occur because brain scans are not conclusive (PCA1.5; C3.10). In one case, the presence of an eye morbidity appears to also confuse the diagnosis (C2.4).

In one case, the couple are referred to a senior specialist in AD and after three visits they get a firm diagnosis (C4.7). Perhaps the couple are referred quickly by their GP because the individual is having particular problems at work and going through an industrial tribunal for inefficiency with difficulties in using a computer, telephone as well as reading and writing, alerting the doctor that something is cognitively wrong (C4.3).

5.3.3 Impact of diagnosis

Once explained, a realisation of why they or their partner have been struggling with their vision begins to form. For some it is relief to know what it is (PCA1.10; PCA2.7), for others, it is overwhelmingly a negative experience (PCA3.6). The importance of having a good explanation of dementia has been highlighted as it can help in understanding the disease and better manage the condition and this seems to have occurred with some of the interviewees (C1.8; C3.12; C4.8; C4.9). One caregiver had not had a carers assessment and poor explanations and lack of empathy causes distress and confusion for others (PCA2.5; PCA2.6; C2.7).

5.4 Theme Three: Loss

The progressive nature of PCA highlights several kinds of losses experienced by the person with PCA. These are: Loss of identity; loss of capabilities; loss of relationships.

5.4.1 Loss of identity

The interviews reveal that the people with PCA are grieving their loss of identity (PCA1.40; PCA2.19; PCA3.6). The loss of the ability to be free and independent is also felt strongly (PCA1.32; PCA2.15; PCA3.6)

Only Laura, whose PCA and tAD symptoms are overlapping, often believes she is still a teacher of children with special needs (C4.32). However, this disorientation is not constant as during our conversation she sometimes appears confused about what she used to do for a living (PCA4.8).

5.4.2 Loss of capabilities

5.4.2.1 Activities of daily living

PCA is causing individuals to lose the ability to undertake many activities of daily living. Lost skills particularly mourned by the interviewees include: inability to prepare food (PCA1.13; PCA2.13; PCA3.10; C3.20); read (PCA1.14; PCA2.12; PCA2.17; PCA3.12; C3.26; C4.19); write (PCA1.14; PCA2.17; C3.29); and understand numbers and money (PCA1.15; C2.16; C3.30; PCA3.15). They are very much aware they struggle over these activities. Frustration (PCA3.6), resignation (PCA1.13) and a fear of looking foolish (PCA3.15) are strongly expressed.

Individuals with PCA want to be able to continue to do things. Some couples have invested in assisted technologies or adaptations for people with visual impairments to help with cooking, baking etc (PCA1.5; C3.18; PCA3.17; C3.31). However, in some cases it appears that carers discourage them to try because of either the length of time it will take for them to do something; the mess they may make whilst doing it (C1.39; C2.12); because they may hurt themselves (C1.11; C2.53) or not succeed in the task (PCA1.24; C1.40) which may upset them or cause agitated behaviours (C1.42; C2.12; C2.48). Carers seem to be 'just managing' most of the time

and any added pressure on them, for instance, taking time to assist their partner, increases their stress levels

Some of the participants of this present study were able to explain their visual difficulties. Their vision is not blurry or unfocused but individual letters or figures as well as lines of text or numbers jump about, so it is impossible to make sense of what they are seeing (PCA1.15; PCA2.20; C3.26).

Interviewees report not being able to identify and differentiate between washing products. This could be because they cannot read the labels on the products, often resorting to colour or smell to identify them (C1.9; C3.15); or because they have lost the ability to recognise familiar objects such as hair shampoo or toothpaste altogether (C2.8; C4.13).

Personal care is a very intimate procedure. To allow somebody, even a close family member, to undertake this role necessitates great trust in that individual. Agitation and aggressive behaviours are reported when caregivers are trying to support their partners with this task (PCA2.8; C4.12).

All four individuals with PCA have difficulty finding their clothes and dressing independently (PCA1.12; C1.10; PCA2.10; C2.9; PCA3.7; C3.16; C3.17; C4.14). Most need a lot of encouragement and support in order to dress themselves and one person has lost this ability totally. They explain their difficulties which include putting clothes on upside down and/or inside out (PCA1.12) and dealing with fasteners such as buttons, hooks and eyes, and zips (C1.10; PCA3.9).

5.4.3 Loss of relationships

A loss or change in the dyadic relationships is reported, particularly by the caregivers. The men of female partners with PCA appear to have felt the changes most. During their working lives, their wives have undertaken all the housekeeping chores and they are now having to learn this in older age, sometimes with disabilities of their own (C1.38); C2.47); C4.37). It is interesting to note that the female caregiver does try and encourage her

partner to do simple household tasks and does not appear to worry about the mess he might make or the time he takes doing them (C3.18). This could be because he is able, although with difficulty, to do some activities at the moment, but his wife's opinion might well change as he progresses through his PCA journey and starts to struggle more.

This change in role can cause friction between couples (PCA2.38). By trying to protect or help their loved ones from being put in harm's way, many of individuals with PCA say they feel frustrated, have their self-confidence diminished or report feeling like they are being treated like a child (PCA2.37; PCA1.41; PCA3.7). Extreme coping behaviours such as locking someone in a room, so they cannot wander off or hurt themselves could be viewed as unethical although the caregiver involved emphasised that he resorted to this because he was so afraid of his loved one getting lost or hurting themselves (PCA2.40).

5.4.4 Caregiver Stressors

In many cases, caregivers describe feeling isolated and being unable to escape from their responsibilities (C1.32; C1.33; C2.56; C3.62; C4.40).

Nevertheless, caregivers seem reluctant to ask or accept support which would enable them to get a break from caring. This may be due to the cost of care (C2.52); not wanting to admit they need help (C2.41); or their belief that the care offered will not be suitable for either their loved one or their own needs (C1.31; C1.32; C2.43; C3.58).

One interview shows the importance of knowing your legal rights as a caregiver is highlighted. One couple is not married and no longer have permission to marry because of dementia. They have not got all their legal affairs in order and it may result in the caregiver losing the family home if their partner is moved into long-term care (C4.38; C4.39)

Another problem that adds stress to caregivers, is challenging behaviours. This can be hurtful remarks (C2.50; C3.62; C4.43) or even aggressive episodes (C4.12; C4.40). None of the caregivers have had access to awareness or educational programmes and find coping with this difficult.

5.5 Theme Four: Navigating the physical environment

Study participants described the difficulties in understanding and navigating the physical environment due to visual perceptual difficulties experienced by the individuals with PCA.

5.5.1 Exercise

Interviewees admit that they did not exercise enough. A lot of the day is spent sitting down watching TV. The people with PCA report that their energy levels are diminished (PCA1.19; PCA2.23; C3.40; C4.23). Perhaps this lack of energy is due to the effort it takes for them to do things because of their visual and/or perceptual inadequacies (PCA1.38)

Brian and Barbara, the youngest of the couples are physically active either walking or swimming regularly but they also experience difficulties as perception problems makes Brian veer to the left often resulting in him walking on the road rather than the pavement increasing his risk of having an accident (PCA3.22; C3.38). The other couples rarely walk or take part in any physical activity (PCA1.19; C1.18; C2.22; C4.23.)

5.5.2 Balance and Falling

The individuals that do not exercise report poor balance issues and a fear of falling (PCA1.11; PCA1.20; C2.23; C4.25). Poor balance appears to be a key deterrent in the people with PCA as they express fears that they are unable to walk safely.

For Brian, continually veering to the left poses a threat to safety. He describes a perceptual phenomenon of something pushing or pulling him (C3.38). Betty also describes something 'pulling her' (PCA1.23) when trying to negotiate steps.

Stairs are described as difficult to manage by the people with PCA. Depth perception problems make coming down stairs particularly problematic (PCA1.23; PCA2.25) and caregivers seem aware of this and endeavour to support their partners when negotiating stairs and uneven surfaces (C2.25; C3.39)

The couples live in various parts of the UK but pavements are consistently called problematic because of their unevenness and are potentially dangerous to walk on if the person with PCA is not holding on to their caregiver (C1.20; C2.24; C4.25).

5.5.3 Visual/Perceptual difficulties

The individuals living with PCA struggle with some similar as well as diverse visual/perceptual difficulties. Most describe not perceiving a whole scene properly, missing objects or people directly in front of them (PCA1.16; PCA2.21; C3.1; C3.3; C3.24; C4.21). Difficulty in using buttons on a telephone is also commonly described (PCA1.25; C3.30; C4.3).

Individuals describe mistaking shapes, shadows or patterns, believing them to be people or objects or even holes in the ground (PCA1.24; PCA3.18; C4.24).

Despite reticence in using stairs, lifts are apt to make people feel even more uncomfortable. Fears are variously described but demonstrate how helpless people can feel if they are not able to be in control of a situation (PCA1.25; PCA2.41).

Brian finds lifts frightening if they are walled with mirrors. The myriad of reflections of himself is disorientating (C3.35). This same problem of mirrored walls in a large toilet had also caused so much disorientation that he was unable to find the exit door and had to be rescued by his wife (PCA3.20). This issue does not seem to affect the other participants, Betty and Polly in particular, looked at mirrors often to check their appearance and, in Betty's case, check she still recognises herself (PCA1.47; PCA2.9).

Computers are also difficult to manipulate, both keyboard operated or iPads (PCA1.37) but this may be because the individuals have not used them to any extent during their working lives, with the exception of Laura (C4.3).

Although individuals with PCA in the study find objects sometimes difficult to recognise, only Polly is experiencing prosopagnosia, often not recognising her husband (C2.18).

5.5.4 Getting Lost

Getting lost or disorientated in both inside and outside environments are major difficulties being experienced by the people with PCA. Individuals are getting lost even in familiar places such as their own homes. This causes great distress because on one hand they become confused as to where they are, but on the other they still have the insight to know that something is happening to their perception that is not letting them understand where they are properly. This dilemma is upsetting for both the person with PCA and their caregiver (PCA1.29; PCA1.30; PCA2.30; C1.22; C2.20; C3.41; C3.42). It appears that changes to the placement of furniture in a room can lead to individuals becoming disoriented. For instance, Polly describes how Joe often moves things around making her feel muddled (PCA2.30; PCA2.31).

The risk of their loved ones getting lost in the outside environment is also a continuous worry for the caregivers. They describe incidents that have resulted in them not letting their partner venture out alone (C2.28; C2.27; C2.28). Only Barbara seems happy for Brian to go out with his friend (who also has Alzheimer's) because she feels they won't go far and neighbours will make sure they get home safely (C3.44).

For those with PCA, the fear of getting lost outside the home has knocked their confidence in going out independently (PCA1.31; PCA2.28). Even to accepting being locked in a room when on holiday so they didn't wander off at night and get lost (PCA2.40). However, being held as a virtual prisoner by their caregiver, even with their best intentions at heart, causes frustration (PCA1.32; PCA2.37).

5.5.5 Design of the physical environment

Each interviewee is asked if they think changes to their physical environment will help them to cope with either their own or their loved one's visual challenges.

Specific colours are not thought of as helpful in improving the home environment as strong contrast (C1.26; C3.18; C3.46). Joe is quite negative when talking about using colour in the home. He doesn't think this will help and is resistant to changing the décor of his home (C2.34).

Light is considered an important issue. Interviewees report difficulties seeing in low light levels (PCA1.3; C2.31; C4.26) but glare and reflective surfaces are also flagged up as problematic (C2.33; C3.45). For instance, surfaces that are highly patterned or shiny are reported as being disorientating and frightening to walk on (PCA1.24; C4.27).

5.6 Theme Five: Social Supports

During the interviews, the couples speak about both negative and positive examples of support.

5.6.1 Family

In all four cases, the key supporter to the person with PCA is their partner. In three of the cases, their children offer some help and assistance of a practical nature (PCA1.15; PCA2.34; C3.53).

Unfortunately, participants also report that some of their family members find it hard to understand that there is anything wrong with the person with PCA. This could be because they do not present initially with symptoms such as memory loss (C4.29; C3.54).

5.6.2 Friends

Out of the four couples, only one couple have managed to continue their close relationships with friends they have known for many years. In their case, although their friendship circle is aging, and all have long term conditions of their own, they continue to support each other though some do not fully understand PCA specific challenges (C1.29; C1.30).

Others have lost touch with friends and are living quite isolated lives relying very much on each other (C2.36; C3.55).

5.6.3 Health and Social Care

Care packages offered differ as the couples all live in different council areas. Only one couple has a comprehensive care package (C4.32). Often the caregivers report unhelpful care staff and inappropriate support because PCA is not understood by health and social care professionals because of their lack of awareness of a person's specific challenges (C1.31; C1.35; C2.43). One caregiver had not had a carers assessment yet (C3.60).

In this study, caregivers state they are keen to have access to a support group, specifically for people with PCA and their caregivers. However, because of the rarity of the condition, they are finding it hard to contact individuals with a similar condition. Unfortunately, the present PCA support group is situated too far away for any of the couples to have regular attendance (C1.37; C4.31).

5.7 Theme Six: Facing the future

Interviewees are asked for their thoughts about the future.

5.7.1 The person with PCA

The people living with PCA thoughts focus on themselves and how PCA might affect them in the future, rather than considering the sort of care they might need to receive as their condition deteriorates (PCA1.48; PCA2.43; PCA3.28).

5.7.2 The Caregiver

On the other hand, the caregivers are much more focused on long term care and its financial impact. These thoughts spark a distressed emotional response (C1.48; C2.57; C3.67; C3.68; C4.45).

5.8 Theme Seven: The emotional impact of living with PCA

Living with dementia impacts on people with dementia and their caregivers on many different levels: emotional, psychological and practical. During the interviews, an array of different emotions regarding the effects PCA is having on individuals continually emerged as they talk about their experiences.

5.8.1 Stoical

The couples speak about their reaction to the diagnosis and what it means to their lives. At a surface level, for some the diagnosis is referred to in a *'matter-of-fact'* manner (PCA1.10; C1.8; PCA2.42).

5.8.2 Despair

On the other hand, others express despair particularly regarding how PCA has impacted on their lives (PCA1.14; PCA2.6; PCA3.6). Caregivers describe their feeling of despair when their loved ones say hateful things to them (C2.18; C4.35).

5.8.3 Fear

Fear appears to be an overriding emotion that the people living with PCA say they often experience. Sometimes they are fearful about how their condition may progress in the future (PCA1.48; PCA2.4; PCA4.10). At other times, fear can engulf them when they find themselves having to cope with a situation that they cannot control or understand (PCA1.22; PCA1.23; PCA1.30; PCA3.2; PCA2.41; PCA3.20). One person seems fearful about how her partner will react if he knows what she is saying to the researcher (PCA2.39).

5.8.4 Frustration

The couples speak about their daily frustrations. For those with PCA, frustration arises from not being able to do the activities and hobbies they used to enjoy (PCA1.14; PCA2.38; PCA3.6).

For the caregivers, different types of frustrations emerge from our conversations. One key frustration appears to be the lack of time for themselves. Their responsibilities mean that they are always in demand, always on duty, often it appears to the detriment of their own health (C3.65; C3.62; C4.40)

Sometimes frustrations arise from the perception that they are not able to get their message across to others who did not understand the condition of PCA (C1.32; C2.46; C4.35).

5.8.5 Anger

Frustration can sometimes spill over into anger. Caregivers express anger about the circumstances they find themselves in (C3.60), or the way they are treated by those in positions of power and authority (C2.7). Sometimes caregivers become angry with themselves because they are finding it difficult to manage with their caring role (C1.46)

5.8.6 Sadness

Individuals living with PCA openly express sadness about having the condition (PCA1.46; PCA2.15). Conversely, caregivers were quite guarded in their emotions but when asked about their feelings during the interviews, the male caregivers personas began to slip, and they became emotionally overwhelmed (C1.47; C2.57).

5.8.7 Lack of confidence

The continuing deterioration of abilities, coupled with caregivers stopping them attempting activities for fear of accidents, have resulted in the individuals living with PCA demonstrating a loss of confidence, explaining they often fear of being thought to be foolish or showing their carer up (PCA1.12; PCA2.41; PCA2.43; PCA3.15; PCA3.16).

5.8.8 Loneliness

The rarity of the condition causes the caregivers to often feel isolate because they are not able to talk to someone who understands the condition of PCA (C1.34; C4.31). They are also aware that their partners are often lonely and need their constant reassurance (C3.55).

5.8.9 Tiredness

All participants talked about struggling and feeling tired a lot of the time. For people with PCA, tiredness came because they had to struggle so much to accomplish tasks and often gave up (PCA1.38; C3.40) and caregivers felt tired due to the care pressures they experienced on a daily basis (C2.47).

5.9 Conclusions

This chapter has used themes to consolidate the views of eight individuals either from the perspective of a person with PCA or a caregiver detailed in [Chapter Four](#) in order to highlight different areas in their lives which they believe are particularly affected by the condition of PCA.

In the next chapter I will compare and contrast these findings with the literature detailed in [Chapter 2](#).

6.0 Chapter Six: How the findings relate to the literature.

6.1 Introduction

This chapter will discuss how the findings from the study relate to the current dementia/PCA literature detailed in [Chapter 2](#). I have used the Theme headings discussed in [Chapter 5](#) to guide the discussion.

6.2 Theme One: Awareness and understanding of PCA

Findings from both the literature and the study confirm that PCA is a rare atypical form of dementia and presents in a different way to tAD with individuals retaining insight and memory but having increasingly reduced vision and perceptual capabilities (Benson, Davis, & Snyder, 1988).

Beh, *et al.* (2015) suggested that the condition is not easily recognised or understood by health and social care professionals or the general public. All four couples not only had difficulties understanding the symptoms themselves but also found health and social care professionals were unaware of PCA.

Currently, the literature shows that PCA is mainly studied by specialist neurologists who are interested in this rare AD variant and most articles appear in academic journals and are medical in content. Researchers are mainly focused on the scientific aspect of how PCA occurs in the brain. There is a paucity of studies exploring how PCA impacts on the social and psychological day to day lives of an individual. It is difficult to get information about the symptoms unless you are aware of PCA in the first place.

6.3 Theme Two: Journey to getting a diagnosis

The literature shows that PCA symptoms, as with most dementias, don't suddenly strike a person but progress insidiously over time with changes in the brain beginning years before there are any signs of the disease (Crutch, et al., 2012). Both the literature and research findings state that PCA initially causes visual and perceptual capabilities to deteriorate in the first

instance rather than memory. (Beh, et al., 2015) have suggested that because of the rarity of the condition, a diagnosis of PCA may be overlooked.

The literature reports that AD usually starts with memory loss rather than visual difficulties (Moore, 2009). Such problems are noticed by family and friends and the first port of call is their GP (Cayton, Graham, & Warner, 2008). This contrasts with people with PCA, where memory, at least in the initial stages, is not problematic (Schott & Crutch, 2019) and both the literature and study findings report that the individual with PCA initiates visits to the opticians thinking their eyesight is faulty. This is understandable as generally people in their 50s and 60s find they need spectacles to read, write and drive at this age (NHS, 2018). Because they are usually younger individuals, often of working age, the visual difficulties are not thought to be due to cognitive causes either by themselves or the optician (Crutch, Walton, Carton, & Shakespeare, 2017).

Study findings show that as their visual and perceptual difficulties deteriorate individuals finally go to see their GP. Because they are aware of their visual and perceptual problems, they can explain what they are experiencing. This insight can lead to stress and worry and indeed, the study findings detail that individuals expressed such emotions. Because individuals were under 65 years and general awareness of the symptoms of PCA is poor, their GPs' put symptoms down to stress and anxiety and prescribed antidepressants in the first instance. The literature states this is often the case in people with PCA, as well as people with other early onset dementias (Draper, et al., 2016). There is a protracted time lapse before diagnosis because of PCAs atypical nature and the fact that brain scans, in the early stages, can be non conclusive (Crutch, 2014).

The literature shows that the impact of a diagnosis of any type of dementia can be traumatic (Górska,, Forsyth, & Maciver, 2018). The importance of having a good explanation of dementia has been highlighted as it can help in understanding the disease and better manage the condition (SCIE, 2015). However, findings from the study revealed that even when a diagnosis has

been made, because many social care staff were unaware of the specific manifestations of PCA, appropriate support for both the person with PCA and their caregiver was lacking.

6.4 Theme Three: Loss

The losses felt by the experience of dementia have been well documented (Górska,, Forsyth, & Maciver, 2018). The literature reports that the lack of ability to undertake many activities of daily living are different in a person with PCA compared to someone with tAD. PCA causes individuals to lose the ability to undertake many activities of daily living at an early stage of the disease (Benson, Davis, & Snyder, 1988). A key contrast between the literature and the findings of the study is that the literature focuses on the scientific reasons for the deterioration of a person's abilities but hardly mentions how these losses affect the person with PCA and their caregiver from a subjective point of view.

The literature reports that reading and writing abilities are commonly affected by PCA (Crutch & Warrington, 2010) and this is echoed in the study findings. Specific losses were felt intensely by the interviewees including: the loss of the role of being a housewife, loss of the ability to read and write, loss of being able to use a credit card; loss of being a driver; loss of the ability to wash and dress independently; whilst their memory appears fine. This differs from someone with tAD who can still perform these daily living activities for quite a long time after initial diagnosis with problems focusing on loss of memory or poor planning skills.

Findings from both the dementia literature as well as the interviews with the study participants show a loss or change in the dyadic relationships (Derksen, Vernooij-Dassen, Gilliss, Olde-Rollert, & Scheltens, 2005). Agitation and aggressive behaviours have been reported when caregivers of people with dementia are trying to support their partners with personal care but this behaviour may be the result of feelings of vulnerability and loss of privacy and dignity (James, 2011). Study findings showed that the individuals with PCA did report that they often felt frustrated and unhappy particularly at not being able to accomplish personal care without help.

Study findings revealed that caregivers were reluctant to ask or accept support which would enable them to get a break from caring even though they often felt tired because of believing that the care offered will not be suitable for either their loved one or their own needs. The literature emphasises the importance of caregivers being signposted to carer support services at the time of diagnosis to help reduce anxieties by equipping them with correct information (Newbrunner, Chamberlain, Borthwick, Baxter, & Glendinning, 2013). The caregivers in this study expressed feelings that the lack of awareness of the day to day challenges caused by PCA meant that the care offered was unhelpful. In addition, a recent survey of caregivers of people with dementia noted that often they were not being offered a carers assessment and that support and respite services were inadequate (Alzheimer's Society, 2020). If this is the case, then supporting someone who has unusual dementia symptoms is understandable.

Findings from the study showed that none of the caregivers have had access to awareness or educational programmes and find coping with this difficult. The value of such training is emphasised in NICE dementia guidelines (NICE, 2016) and might help them cope better when negative incidences arise.

6.5 Theme Four: Navigating the physical environment

Difficulties in understanding and navigating the physical environment have been reported due to visual perceptual difficulties experienced in PCA (Alzheimer's Society, 2018; Crutch, Yong, & Shakespeare, 2016) and this was echoed by the study findings. The importance of having a dementia friendly home was highlighted in the literature and participants appeared content to be living in their home environment although it should be noted that although most of the study had removed patterned carpets, the use of colour contrast and equipment used by those with visual impairments such as talking watches, etc had not been tried by all the couples or offered by social support services. The literature shows that assisted technologies can improve the safety of people with dementia particularly around falls risk and accidents (Brimsa & Oliver, 2019), there

has not been any specific studies focusing on the specific needs of people with PCA.

The literature reports that visually impaired individuals have a significantly greater risk of diminished balance than people with normal vision (Willis, Vitale, Agrawal, & Ramulu, 2013). Poor balance is a key deterrent in the people with PCA walking safely.

Reasons other than having PCA for poor balance were suggested.

Sedentary lifestyles have been shown to lead to muscle weakness leading to difficulties in gait, balance and a significant risk factor for falling.

Findings from this study found that most of the couples admitted they didn't exercise enough and this lack of physical training was having a deteriorating effect on their balance and causing a fear of falling in the individuals with PCA.

The literature reported that individuals living with PCA struggle with a range of diverse visual/perceptual difficulties (Beh, et al., 2015). Study findings described letters and numbers jumping when people were trying to read, mistaking shapes, shadows or patterns sometimes believing them to be people or objects or even holes in the ground. These problems started early on in their PCA journey whereas they normally occur at a later stage in tAD (Crutch, Walton, Carton, & Shakespeare, 2017).

Findings shows that getting lost or disorientated in both inside and outside the home environment were major difficulties being experienced by study participants with PCA. This has been described in the literature by (Tang-Wai & Mapstone, 2006). The risk of their loved ones getting lost in the outside environment is also a continuous worry for the caregivers of people with dementia.

6.6 Theme Five: Social Supports

The support given to a person living with dementia can have a huge impact on their emotional, social and psychological wellbeing by helping them to feel valued and included (Connell, Sanford, & Lewis, 2007). The literature states that the Alzheimer's Society undertook a large initiative to create

'*Dementia Friends*' to change the way people think, act and talk about dementia (Alzheimer's Society, 2018). The stigma of dementia often means that people lose old friendships. Unfortunately, the findings from this study all described the loss of friends as well as family members not believing the person with PCA had a problem. Findings show that support offered by social care services was inadequate with offers of walking frames and respite facilities unsuitable for someone with sensory problems. Lack of awareness meant that the professionals didn't understand the serious visual and perceptual problems the person with PCA was experiencing and react appropriately.

Appropriate care packages can be of vital help to those caring for a person with dementia at home. Findings from the interviews showed that care packages offered differed because the couples all live in different council areas. Only one couple has a comprehensive care package. The caregivers report unhelpful care staff and inappropriate support because PCA is not understood by health and social care professionals.

The literature has suggested that support groups can increase wellbeing for carers because it allows them to connect with likeminded individuals in the same situation (Carers UK, 2014). Findings from this study show there is currently very few groups that support people with PCA although all the caregivers would be eager to attend if there was a group in their area.

6.7 Theme Six: Facing the future

Advanced care planning is advocated by NHS and social care professionals as well as dementia voluntary organisations. This encourages people to think about, discuss and record their wishes for future care for a time when they may not be able to make decisions for themselves (Alzheimer's Society, 2018). Findings from the study revealed that all the caregivers were aware that they needed to plan for the future.

6.8 Theme Seven: The emotional impact of living with PCA

The literature reports that living with dementia impacts on people with dementia and their caregivers on many different levels emotional, psychological and practical (Górska,, Forsyth, & Maciver, 2018); (Holst &

Hallberg, 2003) including negative emotions such as frustration, anger and embarrassment as well as positive feelings such as love and optimism (Caddell & Clare, 2011) (Karlsson, Savenstedt, Axelsson, & Zingmark, 2014) and this is reflected in the findings from the people with PCA. The interviews were structured to allow the participants to show their emotions regarding their personal experiences. Key emotions that echo through all of the interviews are that of sadness and frustration. Because the majority of the participants with PCA had insight into their condition they were very much aware of their deteriorating condition and the fact that they were unable to do many of the activities and hobbies they had previously undertaken. The caregivers expressed sadness as they watched their loved one become progressively disabled and also feelings of frustration that appropriate support was not available to help them cope with the situation. They expressed feelings of loneliness at losing friendships and that family members were often unhelpful.

6.9 Conclusion

This chapter has compared the literature to the findings of the study. The key message from the study is the lack of awareness about PCA by everyone – health and social professionals as well as the general public and this was inhibiting the provision of information and support for both the people with PCA and their caregivers.

Although this study was small and the results cannot be generalised, I believe that taking account of the findings could begin a discussion with PCA experts regarding the development of a good practice guide. The goal would be for this to be widely circulated, to inform everyone who may come in contact with this atypical dementia and assist them in recognising the signs and symptoms of PCA and be able to improve speed of diagnosis and provide appropriate support services.

The next chapter details the first steps towards the possible contents of such a guide which would be aimed at improving the quality of life for people with PCA and their caregivers.

7.0 Chapter Seven: Good Practice Guide

7.1 Introduction

This chapter proposes the development of a good practice guide which would be of value to anyone with PCA or may need to support somebody with this condition.

Figure Four is a diagrammatic map showing basic elements to be considered for inclusion in a best practice care guide for PCA

The four key areas have been identified for consideration for the guide. I have chosen the drivers for change along with suggested interventions because they address the concerns and experiences cited by the case study interviewees as detailed in the Themes chapter (see [Chapter Five](#)).

Figure Five details a flow chart showing the links between the seven themes and the four drivers for change.

Figure 3: Proposed best practice care guide for people with PCA

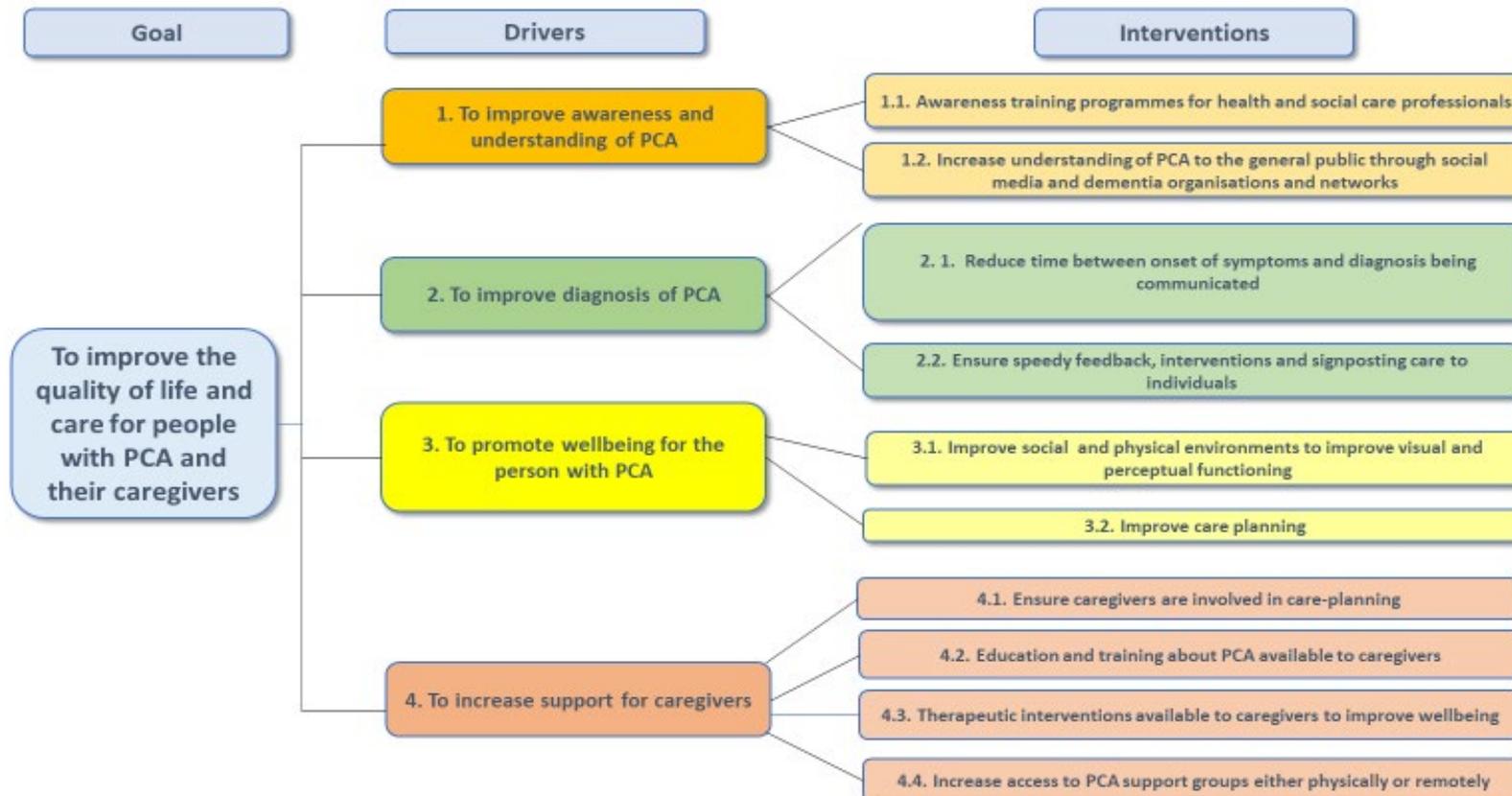
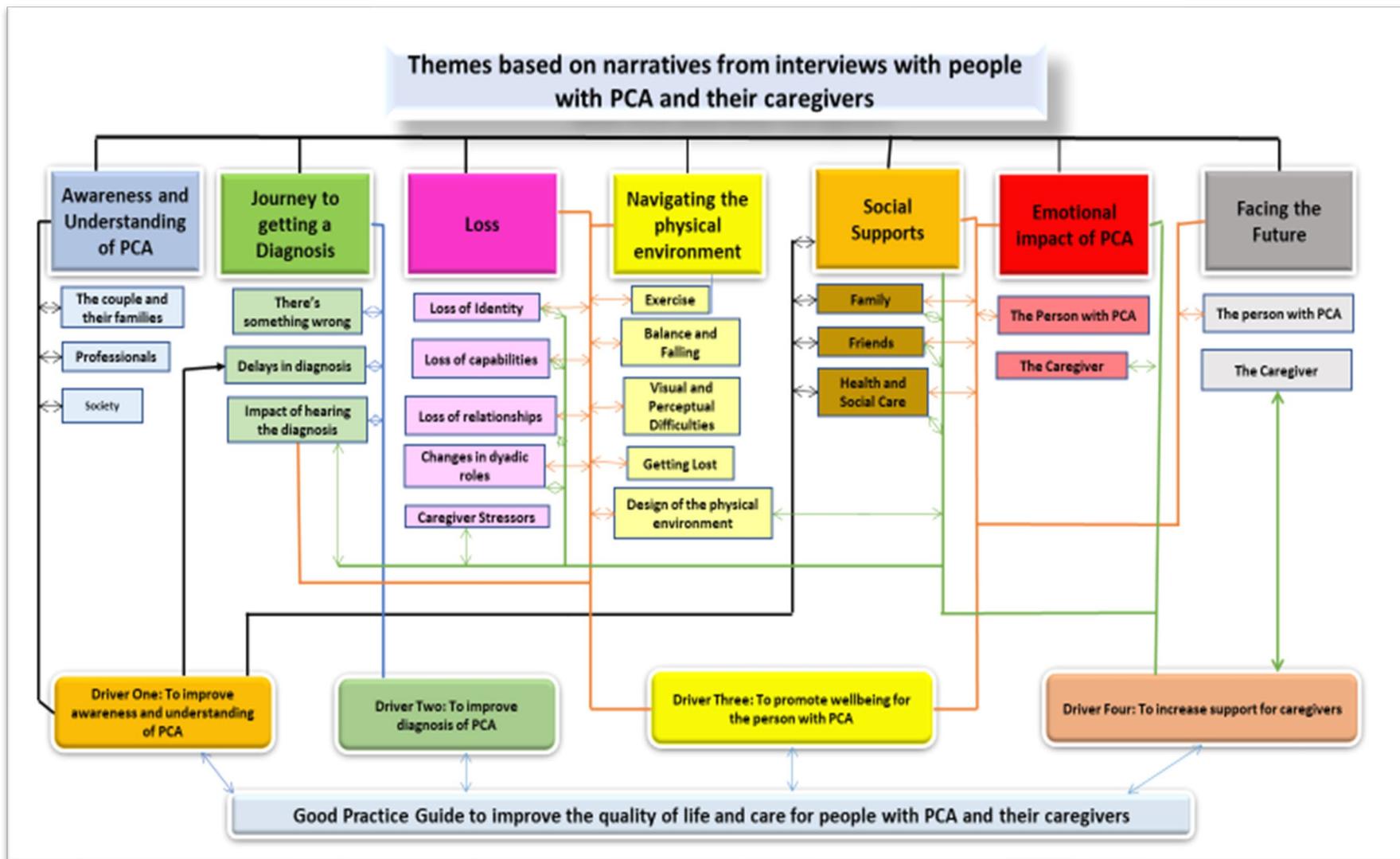


Figure 4: flow chart showing the links between the seven themes and the four drivers for change



7.2 Basic tenets of a Good Practice Guide

The proposed guide would be divided into four parts:

Goal: To improve the quality of life and care for people living with PCA and their caregivers.

Drivers for change: What needs to be delivered to achieve the goal

Interventions: Intentional interaction(s) or event(s) expected to contribute to the successful achievement of the goal

Practical Ways Forward Practical ways in which the interventions can be achieved

Driver 1: To improve awareness and understanding of PCA

Rationale: Improving awareness and understanding of PCA would help to improve diagnosis rates and post-diagnosis service provision.

Why is this needed: Knowledge of PCA is globally lacking by health and social care professionals as well as society in general.

How this could be achieved?

Interventions:

Awareness training for Health and Social Care Professionals

Rationale: Providing skills and resources and encouraging joined-up services between health and social care staff could help people live well with PCA.

Practical ways forward:

Training module added to mainstream courses on dementia for first point of contact health and social care professionals

Comment: The case studies showed that the first point of contact they encountered were not informed about PCA ([Chapter 5, Section 5.2](#)). Indeed, several caregivers took it upon themselves to provide information to the professionals to alert them to the condition of PCA. Therefore, I believe a module about PCA should be added into the syllabus of health and social care professionals who are most likely to encounter someone with PCA. This would include: primary care practitioners; opticians; optometrists; ophthalmologists; neurologists; psychiatrists; psychologists; counsellors; occupational health workers; social care workers; care home staff; community home care workers.

Information packs sent to community health and social care teams

Comment: The case study participants explained that once diagnosed with PCA, support services were uninformed about the condition ([Chapter 5 Section 5.2](#); [Chapter 6 section 6.3 and 6.6](#)). Because community health and social care teams may need to advise and support someone with PCA or their caregivers, information packs containing facts about PCA, its presentation and progression ideally with personal stories from people with the condition, plus contact details of support groups and practical resources that would aid people with visual and cognitive challenges should be provided. This information should be available in written in Plain English, audible and DVD formats and available in a variety of languages.

Roadshows and presentations at health and social care national conferences

Comment: Because of the lack of knowledge about PCA, roadshows and presentations would help to increase knowledge about the condition ([Chapter 5 Section 5.2](#)). However, presentations should be given not only by PCA specialists but also individuals with PCA and their caregivers. This

would ensure a holistic view of the biomedical and psychosocial effects of living with PCA inform audiences.

Increase understanding of PCA to the public

Rationale: Enabling people to recognise the signs and symptoms of PCA will increase awareness of the condition and encourage them to support individuals with PCA living in the community.

Practical ways forward:

Information packs sent to voluntary sector organisations specialising in dementia and/or visual impairment

Comment: The Alzheimer's Society, Alzheimer's Research UK and UCL Rare Dementia Support do have information about PCA on their websites because these organisations are more likely to be contacted by people concerned about visual and/or cognitive difficulties. Individuals from the study had taken it upon themselves to give out information. However, smaller charities such as local visually impaired societies and dementia organisations also need to be informed about this condition. Packs should contain facts about PCA, its presentation and progression ideally with personal stories from people with the condition, plus contact details of support groups and practical resources to aid people with visual and cognitive challenges. Information must be written in plain English format in written, audible and DVD formats and available in a variety of languages.

Information packs available to workplace HR and occupational health teams

Comment: Two people in our study were still in employment when they started with visual and cognitive problems. In one case, the person was undergoing a tribunal for poor performance ([Chapter 5 Section 5.3.2](#)). PCA is an early onset dementia and may affect people who are in employment. It is important that HR personnel and occupational health teams of employers can access information that might be affecting

members of their workforce. Information packs must contain facts about PCA, its presentation and progression ideally with personal stories from people with the condition, plus contact details of support groups and practical resources to aid people with visual and cognitive challenges. Information must be written in plain English format available in written, audible and DVD formats and available in a variety of languages.

Storyline on TV and/or radio

Comment: An effective way of getting a health or social message to millions of people is to highlight a condition using a well-known character within popular TV series. Other formats could include documentaries, theatre productions, podcasts.

Presentations and Posters about PCA at National Dementia Congress

Comment: The National Dementia Congress is an annual event where dementia experts meet to discuss present and future research and care practices for dementia. It is therefore an ideal venue to highlight the needs of people with PCA with presentations or poster presence.

PCA awareness promotion at major public events

Comment: Key public events that gain enormous media coverage are ideal opportunities to highlight PCA. Examples include the London Marathon and Great North Run. Consideration should be given to approaching the Royal Horticultural Society. Every year gardens are created at the Chelsea Flower Show to depict an illness or a specific condition. Currently there is a focus on mental health. A discussion with them sponsoring a Garden depicting the condition of PCA may be successful.

Social media blogs, twitter or Facebook presence

Comment: Blogs, twitter feeds and a PCA Facebook page are up-to-date methods of getting the PCA message to younger age groups.

Driver 2: To improve diagnosis of PCA

Rationale: Timely diagnosis of PCA will improve access to timely and appropriate care and support services

Why this is needed: There are currently delays in diagnosis of PCA causing individuals to remain unsupported by health and social care teams.

How this could be achieved?

Interventions:

Reduce time between onset of symptoms and diagnosis being communicated.

Rationale: Individual concerns caused by delays in accessing appropriate support regarding visual and perception difficulties will be reduced.

Practical ways forward:

Ensure training and information about PCA is given to first point of contact professionals

Comment: As discussed above ([Chapter 5, Section 5.2](#); [Chapter 6 Section 6.2](#)), a module about PCA should be added into the syllabus of health and social care professionals who are most likely to encounter someone with PCA. This would include: primary care practitioners; opticians; optometrists; ophthalmologists; neurologists; psychiatrists; psychologists; counsellors; occupational health workers; social care workers; care home staff; community home care workers.

Individuals can access rapid referral from primary care to specialist services

Comment: The case study participants spoke of the delays they encountered in being referred for specialist care. ([Chapter 5 Section 5.3.2](#); [Chapter 6, Section 6.3](#)). People concerned about their visual and/or cognitive abilities should be referred by their GP for an appointment with ophthalmic and/or neurologist specialist services within six weeks.

Ensure speedy access to multi-disciplinary assessment and diagnosis

Rationale: The person with PCA and their family must be provided with an explanation about their visual and perception difficulties and allow them to access appropriate treatment, advice and support and enable them to plan for their future care needs.

Practical ways forward:

Multi-disciplinary assessment package readily available

Comments: Participants of the case studies talked about having to keep going back for test after test before diagnosis causing anxiety, frustration and stress ([Chapter 5 Section 5.3.2](#)). A person concerned with their visual/cognitive abilities should be offered a one stop shop assessment including.

- ✓ pre-diagnostic assessment counselling
- ✓ vision assessment
- ✓ neuropsychological assessment
- ✓ brain scan
- ✓ physical health screening

Speedy referral, interventions and signposting of care and support services

Comments: Following diagnosis participants described often feeling unsupported ([Chapter 5 Section 5.6.3](#); [Chapter 6 Section 6.3](#)). Once diagnosed, a person with PCA and their caregiver(s) should be offered:

- Referral to post-diagnostic counselling with a therapist trained in PCA
- Access to registration for visual impairment status via referral to ophthalmologist
- Referral to social care services
- An Information pack (audio version available if necessary) which contains:
 - ✓ information about PCA
 - ✓ information about accessing support services
 - ✓ advice regarding applying for financial benefits
 - ✓ information on voluntary care support groups

Driver 3: To promote wellbeing for people with PCA

Rationale: The person with PCA will be able to exercise choice and control over the support they receive and realise their full potential.

Why this is needed: People with PCA need assistance in activities of daily living and can become depressed and isolated through lack of mental and social stimulation.

How this could be achieved?

Interventions:

Improve social and physical environments to aid visual and perceptual functioning

Rationale: People with PCA will be supported to navigate their physical environment and improve social functioning.

Practical ways forward:

Environmental audit of home environment

Comment: Some of the case study participants found their home environment did not support their visual and cognitive needs ([Chapter 5 Section 5.5.5](#); [Chapter 6 Section 6.5](#)). A person with PCA and their caregiver(s) should be offered an environmental audit of their home using dementia design principles to ascertain if their visual and cognitive needs are being met. Occupation health should then liaise with audit team and family to provide appropriate equipment to enable independence plus advise on how to access grants if major building work needs to be undertaken (for example: accessible bathroom; improved lighting).

Assisted technologies to address visual and cognitive needs

Comment: Some, but not all the case study participants had found assisted technology helpful in aiding them to continue daily living activities

[\(Chapter 5 Section 5.4.2.1\)](#). A person with PCA and their caregiver(s) should be offered information about assisted technologies which may help address their visual and cognitive needs plus information about financial assistance to hire or purchase this equipment.

Access to evidence based psychological therapies

Comment: The case study participants showed symptoms of sadness and anxiety [\(Chapter 5 Section 5.8\)](#). It may be helpful if they are offered access evidence based psychological therapies to address mental health issues, if required, but with a therapist who understands the condition of PCA.

The person with PCA has access to a PCA Support Group

Comment: Only one couple in the study had been able to travel to London to attend the PCA support group [\(Chapter 5 Section 5.6.3; Chapter 6 Section 6.6\)](#). The person with PCA should be given information to access a PCA Support Group either physically or remotely. More local groups are needed so access to the support group through Skype or FaceTime links should be explored.

Improve care planning for the person with PCA and their caregiver(s)

Rationale: Empowering individuals with PCA to plan their care will promote independence and choice and enable them to be more involved in decisions about their support needs.

Practical ways forward:

The person with PCA and their caregiver(s) are actively involved in the care planning process

Comment: The case study participants were all interdependent on each other, so it is important that the caregiver(s) are involved with the care planning alongside the person with PCA to ensure that the care plan is achievable by all involved parties [\(Chapter 5 Section 5.6\)](#).

The needs and aspirations of the person with PCA are addressed within the care planning process

Comment: The case study participants who were living with PCA spoke of losing their identity because they could no longer do many of the activities they once enjoyed ([Chapter 5 Section 5.4](#); [Chapter 6 Section 6.4](#)). Of particular note was the lack of physical exercise which was impacting on them maintaining a healthy lifestyle ([Chapter 5 Section 5.5.1](#)). The individual with PCA, with their caregiver(s) should work in partnership with a social care professional to develop a personalised support plan. This should:

- ✓ agree the individual's goals
- ✓ support self-care, including physical exercise and healthy lifestyle
- ✓ document any on-going treatment or medications
- ✓ agree access to local dementia support groups or structured education programme (such as Expert Patient Programme)
- ✓ provide relevant information about PCA support groups (national and regional)

Care and support is provided by health and social care staff who have undergone training and have a good understanding of PCA.

Comment: In order to support a person with PCA it is important that the health and social care staff have undergone appropriate training in order to appropriately assist the person with PCA with the development of their care plan ([Chapter 5 Section 5.6.3](#); [Chapter 6 Section 6.2](#)).

The person with PCA has access to appropriate respite care that meets their needs.

Respite care should be offered to the person with PCA which meets their specific visual and cognitive needs.

Comment: The study showed that often respite, including day care, was often not appropriate for individuals with PCA ([Chapter 6 Section 6.6](#)).

Social care workers need to be aware of their visual and cognitive needs to ensure the placement is successful.

Driver 4: To increase support for caregivers

Rationale: Caregivers will feel supported in their caring role, or if they decide to stop caring.

Why this is needed: Caregivers of people with PCA can suffer from reduced mental and physical health due to feelings of isolation and the pressure of their caring role.

How this could be achieved?

Interventions:

Ensure caregivers are involved in care planning

Rationale: Caregivers can offer a unique perspective through their personal experiences of the person with PCA regarding the type of care that will best support them.

Practical ways forward:

Caregivers are involved in the care planning process

Caregivers will work in partnership with the person with PCA and a social care professional to agree an appropriate support plan.

Comment: As discussed above it is important that the caregiver(s) and the person with PCA work together on the support plan to ensure that it is achievable for both parties.

Caregivers are offered a carers assessment

Caregivers will be offered a carers assessment to address their specific needs and given access to continuing support and advice from their local carers centre.

Comment: Not all the caregivers in the study had had a carers assessment and when they had had, their care needs were not followed through to highlight any problems in addressing them ([Chapter 5, Section 5.4.4](#)). For instance, the caregiver is advised they need time out from their caring, but sometimes circumstances block this happening. For example, the caregiver not being happy about leaving their loved one, or respite not being suitable. Therefore, their need for a break is not being met.

Respite care services are available to caregivers

Respite care services will be offered to caregiver(s), as required, to ensure they have a break from their caring responsibilities

Comment: Respite offered to the caregivers in the study was often not taken up because they didn't think it was appropriate either socially or environmentally ([Chapter 6, Section 6.4](#)). If the person organising respite for the couples had training in the needs of someone with PCA, these issues might be successfully addressed.

Education and training about PCA are available to caregivers

Rationale: Education and training will inform and skill up caregivers to enable them to provide appropriate care and support to a person with PCA.

Practical ways forward:

Caregivers have access to training in group settings and/or online

Comment: None of the caregivers appear to have had any training in how to care for the person they are supporting or addressing their own health needs. They also had to inform themselves about PCA rather than receive this information from health and social care professionals ([Chapter](#)

[5, Section 5.4.4; Chapter 6, Section 6.4](#)). Caregivers must have access to training programmes that give information about PCA, practical problems that can occur when caring (e.g. lifting; challenging behaviours; stress). Training should be offered in a variety of formats including group settings and online. Methods to reduce stress and maintain a healthy lifestyle should be part of the syllabus.

Caregivers can access ongoing training available in a variety of settings, as appropriate

Comment: As with all dementias, changes occur as the disease progresses. It is important that caregivers can access ongoing training and support, as required. This was not happening with the care study participants ([Chapter 5, Section 5.4.4](#)).

Caregivers can access financial and legal advice services

Comment: Caregivers should be able to access financial and legal advice services, if required including being given practical help in applying for financial benefits (if appropriate). The need for this was highlighted by one participant who, because the couple were not married, risked losing the family home if long term care became necessary ([Chapter 5 Section 5.4.4](#)).

Therapeutic interventions are available to caregivers

Rationale: Access to mental health support services will reduce stress and isolation and enable caregivers to continue to care or support them if they need to give up their caring role.

Practical ways forward:

Evidence-based therapies for sadness and loss, including individual and group counselling, is available.

Comment: The caregivers in the study were all suffering from stress and had not seemed to have been offered counselling. One person thought counselling would not be helpful because the counsellor wouldn't understand PCA ([Chapter 5 Section 5.4.4](#)). Caregivers should be able to

access evidence-based psychological therapies for depression and anxiety with a therapist who understands the condition. Sessions should be available in a variety of settings including individual and group counselling, online and via the telephone.

In addition, caregiver(s) must be offered:

- a carers assessment
- a personalised care plan to support their needs
- counselling (if appropriate)
- Access to their local Carer Centre for information and friendship

Increase access to PCA Support Groups, either physically or remotely

Rationale: Access to support groups specifically for PCA carers and family members will provide up-to-date information about the condition as well as practical and emotional support.

Practical ways forward:

Nationwide PCA Support Group access

Comment: The couples were eager to get in contact with other people in the same situation as themselves but only one couple has managed to physically access a support group ([Chapter 5 Section 5.6.3](#); [Chapter 6 Section 6.6](#)). Caregivers need access to national and regional PCA support groups, either physically or remotely. If a local PCA support group does not exist, local councils and dementia charities should be encouraged to provide funding to enable such a support group to be established.

Access to 24/7 support via social media

Comment: Some, but not all individuals in the study were computer literate ([Chapter 5 Section 5.6.3](#); [Chapter 6 Section 6.6](#)). At a national level, social media blogs, chatrooms etc should be set up via PCA Support Groups to allow caregivers remote access to information and support 24/7 but training would need to be given.

7.3 Conclusion

This chapter has detailed the basic tenets of a Good Practice Guide, proposed contents described and practical ways forward to improve wellbeing and support for people with PCA and their caregivers suggested.

The next chapter will review the aim and objectives of my research study and consider if they have been addressed within this document.

8.0 Chapter Eight: Conclusion

8.1 Introduction

This chapter will describe what I did to achieve the objectives of the study and the relevance of this work to current dementia literature. Limitations of the study will be considered and future research and good practice recommendations proposed.

8.2 Overview of the study

The **overall aim** of the study is to explore and understand the everyday experiences of community-dwelling people living with PCA, and their caregivers. This investigation is a necessary precursor to further study around the development of a sensory model of dementia care.

Table 24 is a duplicate of **Table 9** which states the objectives of the study:

Table 24: Objectives of the study

1.	Gain a subjective understanding of the everyday challenges faced by individuals living with PCA
2.	Explore the specific visual needs of community dwelling people living with PCA.
3.	Understand the stressors experienced by family members whilst caring for a person living with PCA.
4.	Identify triggers that could result in the deterioration of the individual wellbeing of people living with PCA.
5.	Consider if the development of a sensory dementia care model would benefit people living in the community who are experiencing dementia and additional sensory problems.

To explore these practical 'life-world problems and issues, I decided to conduct a qualitative investigation using an instrumental multi-case study design. This was a dyadic study. The inclusion criterion was that participants had to be community dwelling couples. One of each pair had a

clinical diagnosis of PCA due to AD and their partner was their main caregiver.

I used purposive sampling which is a deliberate non-random method of sampling. This was so I could identify people with specific characteristics, in this case, the condition of PCA. This is a rare atypical type of dementia and I believed suitable participants would be difficult to locate from a wider dementia population. In order to find people with PCA, I used a gatekeeper from the London PCA Support Group who was able to locate appropriate couples and ask if they would be interested in taking part in my investigations. Four couples agreed to meet with me and participate in the research.

All participants were given an information sheet about the project in order to fully understand the aims and objectives of the study. Because the people with PCA had lost the ability to read, this information was read to them and an offer of a taped version of the sheet given. Participants were able to ask questions about the process and be reassured about the confidentiality of the study.

The consent method I used was '*process consent*'. This procedure entailed continual explanation about the nature and purpose of the study.

Therefore, at the start and throughout the interviews, consent to proceed was sought and participants reassured they could stop the interview at any time without giving a reason. To ensure anonymity, pseudonyms for participants, and other people or places implicated in their accounts were used.

Following two brainstorming sessions with my tutor and medical experts in PCA, a topic guide to assist me with the interviews was developed.

I used a phenomenological approach both to conduct the interviews and at the analysis stage. Because the focus of phenomenology is on the individual, using this form of analysis allows individual stories to be explored and evaluated. When conducting an interview using

phenomenology, it is necessary for the interviewer to 'bracket' off any former knowledge of the subject and personal assumptions they may have about the interviewee. Because I am a caregiver myself, I had to practice removing the feelings I had about my caring position and not assume I knew how that individual was feeling and so I conducted a number of practice interviews in order to feel more comfortable using this approach before I spoke with the study participants.

I used a computer software package called NVIVO when analysing the data. After reading and rereading the text and listening to the audio tapes I gradually moved the findings from the general to the specific introducing themes which I felt informed my original aim and objectives of the study.

8.3 Did I achieve the aim and objectives of the study?

I believe objectives one to four were achieved through knowledge gained from both the background reading for the study and the interviews. I found that academic journal articles were mainly scientific in nature but were highly educational for me in gaining knowledge about **what** happens in the brain when PCA occurs and how it differs from tAD. The interviews with the study participants helped me to gain subjective knowledge about **how** living with PCA affects the social and emotional world of a person with the condition and that of their caregiver.

Objective Five "**Consider if the development of a sensory dementia care model would benefit people with PCA who are living in the community who are experiencing dementia and additional visual problems**" was determined after the first four objectives were completed. Analysis showed that lack of awareness of PCA by health and social care professionals as well as the general public meant that information, care and support appropriate for this population is required.

I believe a widely circulated good practice guide about PCA should be developed and made available to people who may need to support anyone

with this atypical dementia. A proposed guide for consideration is detailed in [Chapter 7](#).

8.4 Relevance of the study

I believe this small study will be an interesting addition to the academic literature because there is a paucity of studies exploring the subjective experience of these individuals and it may encourage larger studies to be undertaken in the future.

8.5 Limitations of the study

This study is a small scale multi case design using only four case studies and undertaken by one research student. Therefore, the research is vulnerable to criticism in relation to the credibility of generalisations of the findings. It is important to remember that each interview was unique, and the comments made by individuals from this study may not be duplicated when tested on a larger group of people with similar characteristics.

The participants taking part in the interviews were aware that they were being recorded. This may have resulted in altered behaviour which can occur when people are aware that they are being observed. It is hoped that this effect was reduced because I tried to gain confidence and trust in the participants through informal discussions prior to the interviews to answer any queries regarding the study.

Another limitation is one of accuracy. Although the conversations were taped and comprehensively transcribed, these transcriptions were not sent to the participants for clarification. This was for two reasons. (1) One participant with PCA was extremely concerned that their partner did not know what she was discussing with me and I needed to reassure her that our conversation would remain totally confidential.

In addition, (2) the people with PCA had lost the ability to read and it would have required somebody to read out the scripts to them who was not their partner to keep everything confidential. Confidentiality and the right to privacy was part of the ethics permission. Due to my unforeseen

suspension of study due to family illness, I was not able to visit the interviewees again to read back our conversation.

It should also be recognised that this type of study requires the researcher to remain detached and impartial. I do not feel it was a limitation to the study in itself because I took great efforts to remove my personal feelings regarding individuals during the interviews and analysis but am aware that as a full-time caregiver myself, it was sometimes difficult not to relate to some of the situations the caregivers mentioned and consider how I might react in similar circumstances.

8.6 Recommendations for future research and good practice

It must be appreciated that this was a very small study of 4 dyads, so no generalisations can be made from the findings. However, the study has revealed details of how people with PCA and their caregivers deal with daily life with this condition and how it impacts on their relationships. This information can be used to start a conversation about how the lives of people with PCA could be better understood and supported.

8.6.1 Research

Larger Qualitative studies

There is currently limited understanding of the subjective experiences of people with PCA and their caregivers. Larger qualitative studies of this population where their voices are heard would further inform researchers and policy makers about the specific needs of this population.

Pilot a PCA Exercise Programme

The lack of exercise was an important issues as sedentary lifestyles have been shown to lead to poorer balance and walking ability with a higher risk of falling. Physical exercise programmes have already been developed separately for people with tAD and blind and visually impaired individuals, research into how these ideas can be incorporated to encourage people

with dementia and sight loss to exercise safely and effectively which in turn may improve the physical and emotional wellbeing of this community.

Assisted Technologies (AT) to aid wayfinding

Research such as how to improve wayfinding, particularly in the home, would not only assist people with PCA but might also help people with tAD who had disorientation problems.

Household and Recreational Assisted Technologies for PCA

Research into whether the development of household and recreational assistive technologies that consider both progressively cognitive and visual/perceptual problems would be of benefit to people with PCA in maintaining activities of daily living.

Designing environments for people with PCA

Projects investigating if the uses of colour and/or contrast and lighting in interior design benefit orientation of space for people with PCA may also be beneficial.

8.6.2 Best Practice Development

Pilot awareness training programme

The development of appropriate training programmes about PCA for professionals and caregivers could help support individuals with the condition.

Pilot Best Practice Care Guide for people with PCA and their caregivers

The development of the suggested best practice care guideline for people with PCA and their caregivers would appear to be a useful addition for supporting people with dementia and their caregivers in health and social care. Further studies using larger numbers of people with PCA, possibly accessed through the PCA support group networks, as well as involving front line health and social care providers would enable a refined edition of the guidance to be developed.

**A qualitative study exploring the specific everyday social and environmental challenges faced by
community dwelling people living with posterior cortical atrophy (PCA)**

Word Count: 71,589

References

- Adams, T. (2008). *Dementia Care Nursing*. (T. Adams, Ed.) Basingstoke: Palgrave MacMillan.
- Ahmed, S., Baker, I., Jusain, M., Thompson, S., Kipps, C., Hornberger, M., . . . Butler, C. R. (2016). Memory Impairment at Initial Clinical Presentation in Posterior Cortical Atrophy. *Journal of Alzheimer's Disease, 52*, 1245-1250. doi:10.3233/JAD-160018
- Allan, K., & Killick, J. (2008). Communication and relationships: an inclusive social world. In M. Downs, & B. Bowers (Eds.), *Excellence in Dementia Care: Research into Practice* (pp. 212-229). Maidenhead: Open University Press.
- Alves, J., Magalhaes, R., Arantes, M., Cruz, S., Goncalves, O. F., & Sampaio, A. (2015). Cognitive rehabilitation in a visual variant of Alzheimer's disease. *22*(1), 73-78. doi: DOI: 10.1080/23279095.2013.831865
- Alzheimer's Association. (2017). *2017 Alzheimer's Disease: Facts and Figures*. Washington, DC: Alzheimer's Association.
- Alzheimer's Knowledge Exchange. (2016). *Shifting Focus: A guide to understanding dementia behaviour*. Ontario: Alzheimer's Society Ontario.
- Alzheimer's Research UK. (2015, September 21st). *One in three people born in 2015 will develop dementia, new analysis shows*. Retrieved February 25, 2018, from Alzheimer's Research UK: <https://www.alzheimersresearchuk.org/one-in-three-2015-develop-dementia/>
- Alzheimer's Research UK. (2018, July). *Numbers of people in the UK*. Retrieved October 2, 2020, from Alzheimer's Research UK/Dementia Statistics Hub: <https://www.dementiastatistics.org/statistics/numbers-of-people-in-the-uk/>

- Alzheimer's Society. (2018). *Dementia Friends*. Retrieved May 23, 2018, from Alzheimer's Society: <https://www.alzheimers.org.uk/get-involved/dementia-friendly-communities/dementia-friends>
- Alzheimer's Research UK. (2019). *Dementia in the Family: The Impact on Carers*. London: Alzheimer's Research UK.
- Alzheimer's Research UK/Rare Dementia Support. (2017). *What is posterior cortical atrophy?* London: Alzheimer's Research UK.
- Alzheimer's Society (a). (2016). *Sight, perception and hallucinations in dementia: Factsheet 527*. London: Alzheimer's Society. Retrieved April 7, 2017, from https://www.alzheimers.org.uk/download/downloads/id/3369/signht_perception_and_hallucinations_in_dementia.pdf
- Alzheimer's Society (b). (2016). *What is frontotemporal dementia (FTD)? Factsheet 404LP*. London: Alzheimer's Society.
- Alzheimer's Society. (2014, November). *What is vascular dementia? Factsheet 402*. Retrieved March 30, 2017, from [alzheimers.org.uk: https://www.alzheimers.org.uk/download/downloads/id/2427/factsheet_what_is_vascular_dementia.pdf](https://www.alzheimers.org.uk/download/downloads/id/2427/factsheet_what_is_vascular_dementia.pdf)
- Alzheimer's Society. (2015). *Exercise and physical activity: Factsheet 529LP*. London: Alzheimer's Society. Retrieved May 23, 2018
- Alzheimer's Society. (2015, March). *Rarer Types of Dementia*. Retrieved April 10th, 2017, from Alzheimer's Society: https://www.alzheimers.org.uk/info/20007/types_of_dementia/108/rarer_types_of_dementia/8
- Alzheimer's Society. (2015, October). *What is young on-set dementia: Factsheet 440LP*. Retrieved October 2nd, 2020, from [Alzheimers.org.uk: https://www.alzheimers.org.uk/sites/default/files/2019-09/factsheet_what_is_young-onset_dementia.pdf](https://www.alzheimers.org.uk/sites/default/files/2019-09/factsheet_what_is_young-onset_dementia.pdf)

- Alzheimer's Society. (2016). *Driving and dementia: Factsheet 439LP*.
London: Alzheimer's Society.
- Alzheimer's Society. (2017). *Assessment, Process and Tests*. Retrieved April 7, 2021, from Alzheimer's Society:
<https://www.alzheimers.org.uk/about-dementia/symptoms-and-diagnosis/diagnosis/assessment-process-tests?documentID=260>
- Alzheimer's Society. (2017). *Posterior cortical atrophy (PCA)*. Retrieved April 30th, 2017, from alzheimers.org.uk:
<http://www.alz.org/dementia/posterior-cortical-atrophy.asp>
- Alzheimer's Society. (2018). *Decision-making, advance care planning and the Mental Capacity Act 2005*. Retrieved April 23, 2018, from Alzheimer's Society:
https://www.alzheimers.org.uk/info/20091/what_we_think/767/decision-making_advance_care_planning_and_the_mental_capacity_act_2005/4
- Alzheimer's Society. (2018). *Dementia Friends*. Retrieved May 23, 2018, from Alzheimer's Society: <https://www.alzheimers.org.uk/get-involved/dementia-friendly-communities/dementia-friends>
- Alzheimer's Society. (2020). *The Fog of Support: An inquiring into the provision of respite care and carers assessment for people affected by dementia*. London: Alzheimer's Society UK.
- Amis, M. (2001, December 21). *Age Will Win*. Retrieved April 24, 2018, from TheGuardian.com:
<https://www.theguardian.com/film/2001/dec/21/artsfeatures.fiction>
- Andersen, R. A., Anderson, K. N., Hwang, E., & Huaschild, M. (2014). Optic ataxia: from Balint's syndrome to the parietal reach region. *Neuron*, 81(5), 967-983. doi:10.1016/j.neuron.2014.02.025

Andrade, K., Samri, D., Sarazin, M., de Souza, L., Cohen, L., & et al. (2010).
Visual neglect in posterior cortical atrophy. *BMC
Neurology*201010:68, 10(68). doi:[https://doi.org/10.1186/1471-
2377-10-68](https://doi.org/10.1186/1471-2377-10-68)

Aresi, A., & Giovagnoli, A. R. (2009). The Role of Neuropsychology in
Distinguishing the Posterior Cortical Atrophy Syndrome and
Alzheimer's Disease. *Journal of Alzheimer's Disease*(18), 65-70.
doi:10.3233/JAD-2009-1123

Association of British Dispensing Opticians. (2018, February 28th).
Glaucoma. Retrieved February 28th, 2018, from Association of
British Dispensing Opticians: Eyecare FAQ:
[https://www.abdo.org.uk/information-for-the-public/eyecare-
faq/glaucoma/](https://www.abdo.org.uk/information-for-the-public/eyecare-faq/glaucoma/)

Bahar-Fuchs, A., Martyr, A., Goh, M., Sabates, J., & Clare, L. (2019).
Cognitive Training for people with mild to moderate dementia
(Intervention Review). *Cochrane Database of Systematic
Reviews*(3). doi:DOI: 10.1002/14651858.CD013069.pub2

Baltes, P. B., & Lindenberger, U. (1997). Emergence of a powerful
connection between sensory and cognitive functions across the
adult life span: a new window to the study of cognitive aging?
Psychology and Aging, 12(1), 12 - 21.

Bartlett, R., & McKeefry, D. (2009). *People with dementia and sight loss: a
scoping study of models of care*. London: Thomas Pocklington
Trust.

BBC. (2007, July 12). *Granddad, Dementia and Me*. Retrieved April 24,
2018, from YouTube:
<https://www.youtube.com/watch?v=nmfad3dmXSE>

Beh, S. C., Muthusamy, B., Calabresi, P., Hart, J., Zee, D., Patel, V., &
Frohman, E. (2015). Hiding in plain sight: a closer look at posterior

cortical atrophy. *Practical Neurology*, 15(5), 5-13.

doi:10.1136/practneurol-2014-000883

Benson, D. F., Davis, R. J., & Snyder, B. D. (1988). posterior cortical atrophy. *Archives of Neurology*, 45, 789-93.

Bier, N., El-Samra, A., Bottari, C., Vallet, G. T., Carignan, M., Paquette, G., . .
. Rouleau, I. (2019). Posterior cortical atrophy: Impact on daily
living activities and exploration of a cognitive rehabilitation
approach. *Cogent Psychology*, 6.

doi:10.1080/23311908/2019.1634911

Biran, I., & Coslett, H. B. (2003). Visual Agnosia. *Current Neurology and
Neuroscience Report*(3), 508-512. Retrieved July 2, 2016

Bokde, A. L., Pietrini, P., Ibanez, V., & et al. (2001). The effect of brain
atrophy on cerebral hypometabolism in the visual variant of
Alzheimer disease. *Archives of Neurology*, 58, 480-486.

Borruat, F. X. (2013). Posterior Cortical Atrophy: Review of the Recent
Literature. *Current Neurology and Neuroscience Reports*, 13, 406-
413. doi:10.1007/s11910-013-0406-8

Boucart, M., Calais, G., Lenoble, Q., Moroni, C., & Pasquier, F. (2014, July
25). Differential processing of natural scenes in posterior cortical
atrophy and in Alzheimer's disease, as measured with a saccade
choice task. *Frontiers in Integrative Neuroscience*, 8(Article 60).

doi:10.3389/fnint.2014.00060

Bowes, A., McCabe, L., Dawson, A., & Greasley-Adams, C. (2014). *Design of
homes and living spaces for people with dementia and sight loss*.
London: Thomas Pocklington Trust.

Bowling, A. (1997). *Research methods in health: investigating health and
health services*. Buckingham: Open University Press.

- Brawley, E. C. (2001). Environmental design for Alzheimer's disease: a quality of life issue. *Aging and Mental Health*(Supplement 1), S79 - S83.
- Breen, N., Caine, D., & Coltheart, M. (2001). Mirrored-self misidentification: Two cases of focal onset dementia". *Neurocase*. 7: 239–254.
doi:10.1093/neucas/7.3.254. *Neurocase*, 239–254.
doi:10.1093/neucas/7.3.254
- Brimsa, L., & Oliver, K. (2019). Effectiveness of assistive technology in improving the safety of people with dementia: a systematic review and meta-analysis. *Aging and Mental Health*, 23(8), 942-951.
Retrieved from <https://doi.org/10.1080/13607863.2018.1455805>
- Broadaty, H., Seeher, K., & Gibson, L. (2012). Dementia time to death: a systematic literature review on survival time and years of life lost in people with dementia. *International Psychogeriatrics*, 24(7), 1034-1045. doi:10.1017/S1041610211002924
- Brooker, D. (2007). *Person-centred dementia care: making services better*. London: Jessica Kingsley.
- Bryden, C. (2005). *Dancing with Dementia: My story of living positively with dementia*. London: Jessica Kingsley Publishers.
- Buch, H., Vinding, T., & Neilsen, N. V. (2001). Prevalence and causes of visual impairment according to World Health Organisation and United States criteria in an aged, urban Scandinavian population: the Copenhagen City Eye Study. *Ophthalmology*, 108(12), 2347 - 2357.
- Bunn, F., Burn, A.-M., Goodman, C., Rait, G., Norton, S., Robinson, L., . . . Brayne, C. (2014, October 31). Comorbidity and Dementia: a scoping review of the literature. *BMC Medicine*, 12, 192.
doi:10.1186/s12916-014-0192-4

- Burns, A., & Buckman, L. (2013). *Timely diagnosis of dementia: Integrating Perspectives, Achieving Consensus*. London: NHS England/BMA.
- Caddell, L. S., & Clare, L. (2011). I'm still the same person: The impact of early-stage dementia on identity. *Dementia, 10*(3), 379-398.
doi:10.1177%2F1471301211408255
- Cahill, S., Pierce, M., Werner, P., Darley, A., & Bobersky, A. (2015). A Systematic Review of the Public's Knowledge and Understanding of Alzheimer's Disease and Dementia. *Alzheimer Disease and Associated Disorders, 29*, 255-275.
- Caine, D. (2004). Posterior Cortical Atrophy: A Review of the Literature. *Neurocase, 10*(5), 382 - 385. Retrieved July 19, 2011, from <http://dx.doi.org/10.1080/13554790490892239>
- Calkin, M. P. (1997). A Supportive Environment for People with Late-Stage Dementia. In C. R. Kovach (Ed.), *Late Stage Dementia Care: A Basic Guide* (pp. 1001 - 1111). Washington DC: Taylor and Francis.
- Calkins, M. (2002). How colour throws light on design in dementia care. *Journal of Dementia Care, 10*(4), 20 - 23.
- Campbell, S. (2005). *Deteriorating vision, falls and older people: the links*. Glasgow: Visability.
- Cantley, C., Woodhouse, J., & Smith, M. (2005). *Listen to us: involving people with dementia in planning and developing services*. Newcastle: Northumbria University.
- Carers UK. (2014). *Get Support*. Retrieved May 23, 2018, from Carers UK: <https://www.carersuk.org/help-and-advice/get-support>
- Cassim, R. (2007). The ageing eye: most people's eyes change as they age. *Continuing Medical Education, 484* - 488.
- Cayton, H., Graham, N., & Warner, J. (2008). *Alzheimer's and other dementias* (3rd ed.). London: Class Publishing.

- Cedrone, C., Culasso, F., Cesareo, M., Nucci, C., & al, e. (2003). Incidence of blindness and low vision in a sample population: the Priverno Eye Study, Italy. *Ophthalmology*, *110*(3), 584 - 588.
- Chan, D., Crutch, S. J., & Warrington, E. K. (2001). A disorder of colour perception associated with abnormal colour after-images: a defect of the primary visual cortex. *Journal of Neurological Neurosurgery Psychiatry*(71), 515-517. Retrieved July 10, 2011
- Chandra, S. R., & Issac, T. (2014, September). Neurodegeneration and Mirror Image Agnosia. *North American Journal of Medical Sciences*, *6*(9), 472-477. doi:10.4103/19472714.141647
- Chang, U. P., Lorenz, R. A., Phillips, M., Peng, H. L., & Szigeti, K. (2020). Fatigue in Family Caregivers of Individuals with Dementia: Associations between sleep, depression and care recipients functionality. *Journal of Gerontological Nursing*, *46*(9), 14-18. Retrieved April 21, 2021, from <https://doi.org/10.3928/00989134-20200527-01>
- Charles, R. F., & Hillis, A. E. (2005). Posterior cortical atrophy: Clinical presentation and cognitive deficits compared to Alzheimer's disease. *Behavioural Neurology*, *16*, 15-23. Retrieved November 11, 2019
- Chechlac, M., & Humphreys, G. W. (2014, March 7). The enigma of Balint's syndrome: neural substrates and cognitive deficits. *Frontiers in Human Neuroscience*, *8*(Article 123). doi:10.3389/fnhum.2014.00123
- Chester, R., & Bender, M. (1999). *Understanding dementia. The man with the worried eyes*. London: Jessica Kingsley.

- Clare, L. (2003). Managing threats to self: awareness in early stage Alzheimer's disease. *Social Science & Medicine*, *57*, 1017-1029.
doi:10.1016/S0277-9536(02)00476-8
- Collins Hunt, D. (2011). Young-Onset Dementia: A Review of the Literature and What It Means for Clinicians. *Journal of Psychosocial Nursing and Mental Health Services*, *49(4)*, 28-33., *29(4)*, 28-33.
doi:doi.org/10.3928/02793695-20110302-05
- Connell, B. R., Sanford, J. A., & Lewis, D. (2007). Therapeutic effects of an outdoor activity program on nursing home residents with dementia. *Journal of Housing for the Elderly*, *21(3)*, 194-209
- Crews, J. E., & Whittington, F. J. (Eds.). (2000). *Vision Loss in an aging society*. New York: AFB Press.
- Cronin-Golomb, A., & Hof, P. R. (Eds.). (2004). *Vision in Alzheimer's Disease* (Vol. 34). New York: Karger.
- Crutch, S. J. (2014). Seeing why they cannot see: Understanding the syndrome and causes of posterior cortical atrophy. *Journal of Neuropsychology*, *8*, 157-170. doi:10.1111/jnp.12011
- Crutch, S. J., & Warrington, E. K. (2010). The relationship between visual crowding and letter confusability: Towards an understanding of dyslexia in posterior cortical atrophy. *Cognitive Neuropsychology*, *26(5)*, 471-498.
doi:http://dx.doi.org/10.1080/02643290903465819
- Crutch, S. J., Lehmann, M., Gorgoraptis, N., Kaski, D., Ryan, N., Hasai, M., & Warrington, E. K. (2011). Abnormal visual phenomena in posterior cortical atrophy. *Neurocase*, *17(2)*, 160-177.
doi:10.1080/13554794.2010.504729
- Crutch, S. J., Lehmann, M., Schott, J. M., Rabinovici, G. D., Rossor, M. N., & Fox, N. C. (2012). Posterior cortical atrophy. *Lancet Neurology*, *11*,

170-178. Retrieved July 2, 2016, from
www.thelancet.com/neurology

Crutch, S. J., Schott, J. M., Rabinovici, G. D., Boeve, B. F., Cappa, S. F.,
Dickerson, B. C., . . . Fox, N. C. (2013). Shining a light on posterior
cortical atrophy. *Alzheimer's & Dementia*, *9*, 463-465.
doi:org/10.1016/j.jalz.2012.11.004

Crutch, S. J., Schott, J. M., Rabinovici, G. D., Murray, M., Snowden, J. S., van
der Flier, W. M., . . . Fox, N. C. (2017, March 2nd). Consensus
classification of posterior cortical atrophy. *Alzheimer's & Dementia*.
doi:10.1016/j.jalz.2017.01.014

Crutch, S. J., Yong, K. X., & Shakespeare, T. J. (2016). Looking but Not
Seeing: Recent Perspectives on Posterior Cortical Atrophy. *Current
Directions in Psychological Science*, *25*(4), 251-260.
doi:10.1177/096372141665599

Crutch, S., Walton, J., Carton, A., & Shakespeare, T. (2017). *The Stages of
Posterior Cortical Atrophy (PCA)*. Retrieved from
Raredementiasupport.org:
[http://www.raredementiasupport.org/pca/what-is-pca/stages-of-
pca/](http://www.raredementiasupport.org/pca/what-is-pca/stages-of-pca/)

Dalrymple, K. A., Barton, J. J., & Kingstone, A. (2013, April 17). A world
unglued: simultanagnosia as a spatial restriction of attention.
Frontiers in Human Neuroscience, *7*(Article 145).
doi:10.3389/fnhum.2013.00145

Davis, A. S., Trotter, J. S., Hertz, J., Bell, C. D., & Dean, R. S. (2012). Finger
Agnosia and Cognitive Deficits in Patients with Alzheimer's Disease.
Applied Neuropsychology: Adult, *19*(2), 116-120.
doi:10.1080/09084282.2011.6439

Davis, R. (1989). *My Journey into Alzheimer's Disease: Helpful Insights for
Family and Friends*. Illinois: Tyndale House.

- Day, K., Carreon, D., & Stump, C. (2000). The therapeutic design of environments for people with dementia: A review of the empirical research. *The Gerontologist*, 40(4), 397 - 416.
doi:10.1093/geront/40.4.397
- Dementia Services Development Centre. (2010). *10 Helpful Hints for Dementia Design at Home*. Stirling: University of Stirling.
- Dementia UK. (2018). *What is Dementia*. London: Dementia UK. Retrieved November 2, 2020
- Denscombe, M. (2014). *The good research guide for small-scale social research project* (5th ed.). Buckingham: Open University Press.
- Department of Health (a). (2015, May 8). 2010 to 2015 Government policy: Dementia Update. London, UK. Retrieved March 29th, 2017, from <https://www.gov.uk/government/publications/2010-to-2015-government-policy-dementia/2010-to-2015-government-policy-dementia>
- Department of Health (b). (2015). *Prime Minister's Challenge on dementia 2020*. London: Department of Health.
- Department of Health. (2009). *Living with Dementia: A National Dementia Strategy*. London: Crown Copyright.
- Department of Health. (2010). *Quality outcomes for people with dementia: building on the work of the National Dementia Strategy*. Leeds: Department of Health.
- Department of Health. (2013). *Dementia: A state of the nation report on dementia care and support in England*. London: Department of Health.
- Department of Health. (2016). *NHS Outcomes Framework: at-a-glance*. London: Department of Health.

- Department of Health and Social Care. (2016, November 2). *Listening to people with dementia and their carers*. Retrieved March 4, 2018, from Government/News:
<https://www.gov.uk/government/news/listening-to-people-with-dementia-and-their-carers>
- Derksen, E., Vernooij-Dassen, M., Gilliss, F., Olde-Rollert, M., & Scheltens, P. (2005). The impact of diagnostic disclosure in dementia: a qualitative case analysis. *International Psychogeriatrics*, *17*(2), 319-32. doi:0.1017/S1041610205001316
- DH/SCLGCP/SCP/OP&D. (2010). *Quality outcomes for people with dementia: building on the work of the National Dementia Strategy*. Leeds: Department of Health.
- Draper, B., Cations, M., White, F., Trollor, J., Loy, C., Brodiaty, H., . . . Withall, A. (2016). Time to diagnosis in young-onset dementia and its determinants: the INSPIRED study. *International Journal of Geriatric Psychiatry*, *31*(11), 1217-1224. doi:10.1002/gps.4430
- DSDC. (2013). *Improving the design of housing to assist people with dementia*. Stirling: Dementia Services Development Centre.
- Dunne, T. (2004). Improving performance on activities of daily living in Alzheimer's Disease: Practical applications of Vision Research. In A. Cronin-Golomb, & P. R. Hof (Eds.), *Vision in Alzheimer Disease* (Vol. 34, pp. 305 - 324). Basel: Karger.
- Edhlund, B. M., & McDougall, A. G. (2016). *NVivo 11 Essentials*. Stallarholmen, Sweden: Form and Kunskap.
- Ellison, J. M. (2016, November 14th). *A Form of Dementia that Affects Vision: Posterior cortical Atrophy*. Retrieved April 30th, 2017, from brightfocus.org:
<http://www.brightfocus.org/alzheimers/article/form-dementia-affects-vision-posterior-cortical-atrophy>

- Evans, J. R., Fletcher, A. E., & Wormald, R. P. (2004). Causes of visual impairment in people aged 75 years and older in Britain. An add on study to the MRC Trial of Assessment and Management of Older People in the Community. *British Journal of Ophthalmology*, *88*(3), 365-370.
- Evans, J. R., Fletcher, A. E., Wormald, R. P., Siu-Woon Ng, E., Stirling, S., Smeeth, L., . . . Tulloch, A. (2002). Prevalence of visual impairment in people aged 75 years and older in Britain. *Ophthalmology*, *86*, 795 - 800.
- Feil, D. G., Lukman, R., Simon, B., Walston, A., & Vickrey, B. (2011). Impact of dementia on caring for patients' diabetes. *Aging and Mental Health*, *15*(7), 894 - 903.
- Feng-Ping, L., Kun-Pei, L., & Hsu-Ko, K. (2009, January 7). Diabetes and the Risk of Multi-System Aging Phenotypes: A Systematic Review and Meta-Analysis. *PLoS ONE* *4*(1), 4(1).
doi:10.1371/journal.pone.0004144
- Fleming, R., Crookes, P., & Sum, S. (2007). *A Review of the Empirical Literature on the Design of Physical Environments for People with Dementia*. Stirling: Dementia Services Development Centre.
- Fukuoka H, N. M. (2015, October 8th). The occurrence of visual and cognitive impairment, and eye diseases in the super-elderly in Japan: a cross-sectional single-center study. *BMC Research Notes*, *6*19. doi:10.1186/s13104-015-1625-7.
- Ganguli, M., Dodge, H. H., Shen, C., Pandav, R. S., Pandav, R. S., & DeKosky, S. T. (2005). Alzheimer Disease and Mortality. *Archives of Neurology*, *62*, 779-784. Retrieved February 26, 2018, from <https://jamanetwork-com.libproxy.ucl.ac.uk/journals/jamaneurology/fullarticle/788478>

- Gardini, S., Concari, L., Pagliara, S., Ghetti, C., Venneri, A., & Caffarra, P. (2011). Visuo-spatial imagery impairment in posterior cortical atrophy: A cognitive and SPECT study. *Behavioural Neurology, 24*, 123-132. doi:10.3233/BEN-2011-0279
- Gersztenkorn, D., & Lee, A. G. (2014). Palinopsia revamped: A systematic review of the literature. *Survey of Ophthalmology, 60*, 1-35. doi:http://dx.doi.org/10.1016/j.survophthal.2014.06.003
- Giorgi, A. (2009). *The Descriptive Phenomenological Method in Psychology: A Modified Husserlian Approach*. Pittsburgh, Pennsylvania: Duquesne University Press.
- Giorgi, A. (2018). *Reflections on Certain Qualitative and Phenomenological Psychological Methods*. Colorado Springs, CO: University Professors Press.
- Giovagnoli, A. R., Aresi, A., Reati, F., Riva, A., Gobbo, C., & Bizzi, A. (2009, April). The neuropsychological and neuroradiological correlates of slowly progressive visual agnosia. *Neurological Sciences, 30*(2), 123-131. doi:https://doi.org/10.1007/s10072-009-0019-9
- Glaucoma Research Foundation. (2016, November 18). *Glaucoma Facts and Stats*. Retrieved April 7, 2017, from Glaucoma Research Foundation: <http://www.glaucoma.org/glaucoma/glaucoma-facts-and-stats.php>
- Glaucoma Research Foundation. (2017, October 29). *Glaucoma Facts and Stats*. Retrieved February 28, 2018, from Glaucoma Research Foundation: <https://www.glaucoma.org/glaucoma/glaucoma-facts-and-stats.php>
- Goethals, M., & Santens, P. (2001). Posterior cortical atrophy. Two case reports and a review of the literature. *Clinical Neurology and Neurosurgery*(103), 115-119. Retrieved July 1, 2016, from www.elsevier.com/locate/clineuro

- Goldsmith, M. (1996). *Hearing the voice of people with dementia*. London: Jessica Kingsley Publishers.
- Goldstein, M. A., Ivanov, I., & Silverman, M. E. (2011). Posterior cortical atrophy: an exemplar for renovating diagnostic formulation in neuropsychiatry. *Comprehensive Psychiatry*, *52*, 326-333. Retrieved November 11, 2019
- Gonzalez, A. S., Hoffman, P., & Crutch, S. (2019). Where words meet numbers: Comprehension of measurement unit terms in posterior cortical atrophy. *Neuropsychologia*, *131*, 216-222. doi:10.1016/j.neuropsychologia.2019.05.004
- Goodman, C., & Watson, L. (2010). *Design guidance for people with dementia and for people with sight loss: Research Findings Number 35, December 2010*. London: Thomas Pocklington Trust.
- Górska,, S., Forsyth, K., & Maciver, D. (2018). Living With Dementia: A Meta-synthesis of Qualitative Research on the Lived Experience. *Gerontologist*, *58*(3), e180–e196. doi:10.1093/geront/gnw195
- Grand, J., & Feldman, H. H. (2007). Historical concepts of Alzheimer's disease and dementia. In H. H. Feldman (Ed.), *Atlas of Alzheimer's Disease* (pp. 1 - 26). London: Informa Healthcare.
- Greasley-Adams, C., Bowes, A., Dawson, A., & McCabe, L. (2016). *Good practice in the design of homes and living spaces for people with dementia and sight loss*. London: Thomas Pocklington Trust/University of Stirling.
- Harding, E., Sullivan, M. P., Woodbridge, R., Yong, K. X., McIntyre, A., Gilhooly, M. L., . . . Crutch, S. J. (2018). 'Because my brain isn't as active as it should be, my eyes don't always see': a qualitative exploration of the stress process for those living with posterior cortical atrophy. *BMJ Open*. doi:10.1136/bmjopen-2017-018663

- Heber, I. A., Costa, A. S., Werner, C. J., Schone, U., Reich, A., Schulz, J. B., & Reetz, K. (2016, July-September). Posterior Cortical Atrophy: A Case Report of a 6-Year Natural Progression. *Alzheimer Disease and Associated Disorders*, *30*, 276-280.
- Hinds, S. (2021). *Spotlight on PCA*. Retrieved from Dementiacarers.org.uk: <https://dementiacarers.org.uk/virtual-carers-centre/bitesize-learning/spotlight-on-pca/>
- Holroyd, S. (2004). Visual Hallucinations in Alzheimer's Disease. In A. Cronin-Golomb, & P. R. Hof (Eds.), *Vision in Alzheimer's Disease* (pp. 126 - 135). Basel: Karger.
- Holst, G., & Hallberg, I. R. (2003). Exploring the meaning of everyday life, for those suffering from dementia. *American Journal of Alzheimer's Disease and Other Dementia*, *18*(6), 359-65.
- Hughes, J. (Ed.). (2006). *Palliative Care in Severe Dementia*. London: MA Healthcare Limited.
- Hughes, J. C. (2011). *Alzheimer's and other Dementias*. Oxford: Oxford University Press.
- Husain, M., & Stein, J. (1988). Reszo Balint and his most celebrated case. *Archives of Neurology*, *45*(1), 89-93.
doi:0.1001/archneur.1988.00520250095029
- Husserl, E. (1970a (Original work published in 1900)). *Logical investigations*. New York: Humanities Press.
- Husserl, E. (1970b (original work published in 1954)). *The Crisis of European Sciences and Transcendental Phenomenology*. (D. Carr, Trans.) Evanston, IL: Northwestern University Press.
- Husserl, E. (1983 (original work published in 1913)). *Ideas pertaining to a pure phenomenology and to a phenomenological philosophy*. (F. Kersten, Trans.) The Hague: Martinus Nijhoff.

- IGA. (2012). *What is glaucoma*. Retrieved January 7th, 2012, from The International Glaucoma Association: <http://www.glaucoma-association.com/the-eye-and-glaucoma/what-is-glaucoma-.html>
- Isella, V., Villa, G., Mapelli, C., Ferri, F., Appollonio, M., & Ferrarese, C. (2014, October 20). The Neuropsychiatric Profile of Posterior Cortical Atrophy. *Journal of Geriatric Psychiatry and Neurology*, 1-9. doi:10.1177/0891988714554713
- Jahoda, M. (1958). *Current concepts of Positive Mental Health*. New York: Basic Books.
- Jebelli, J. (2017). *In Pursuit of Memory: The Fight against Alzheimer's*. London: John Murray (Publishers).
- Jefferis, J. M., Mosimann, U. P., & Clarke, M. P. (2011). Cataract and Cognitive Impairment: a review. *British Journal of Ophthalmology*, 95, 17 - 23.
- Jolley, D. (2010). An introduction to the dementias: a clinical view. In J. C. Hughes, M. Lloyd-Williams, & G. A. Sachs (Eds.), *Supportive Care for the Person with Dementia* (pp. 11-20). Oxford: Oxford University Press.
- Jones, G., van der Eerden-Rebel, W., & Harding, J. (2006). Visuo-perceptual-cognitive deficits in Alzheimer's disease: adapting a dementia unit. In B. M. Miesen, & G. M. Jones (Eds.), *Care Giving in dementia: research and applications* (Vol. 4, pp. 3-57). London: Routledge.
- Josephs, K. A., Whitwell, J. L., Boeve, B. F., Knopman, D., Knopman, D. S., Tang-Wai, D. F., . . . Petersen, R. C. (2006). Visual Hallucinations in Posterior Cortical Atrophy. *Archives of Neurology* 2006;63:1427-1432(63), 1427-1432. Retrieved July 2, 2016
- Kabel, S. (2001). *The Person Behind Dementia*. Aalborg: Formidlingscentre Nord.

- Karlsson, E., S€avenstedt, S., Axelsson, K., & Zingmark, K. (2014). Stories about life narrated by people with Alzheimer's disease. *Journal of Advanced Nursing*, *70*(12), 2791 -2799. doi:10.1111/jan.12429
- Kas, A., Cruz de Souza, L., Samri, D., Bartolomeo, P., Lacomblez, L., Kalafat, M., . . . Sarazin, M. (2011). Neural correlates of cognitive impairment in Posterior Cortical Atrophy. *Brain*, *134*, 1464-1478. doi:10.1093/brain/awr055
- Keenan, T. D., Goldacre, R., & Goldacre, M. (2014, January). *Associations between Age-Related Macular Degeneration, AlzheimerDisease and Dementia: Record Linkage Study of Hospital Admissions*. Retrieved April 3, 2017, from jamanetwork.com:
<http://jamanetwork.com/journals/jamaophthalmology/fullarticle/1774028>
- Kessing, L. V., Lopez, A. G., Anderson, P. K., & Kessing, S. V. (2007, January). No increased risk of developing Alzheimer disease in patients with glaucoma. *Journal of Glaucoma*, *16*(1), 47-51. Retrieved April 7th, 2017
- Kessler, L. (2007). *Dancing with Rose: finding life in the land of alzheimer's*. London: Viking.
- Khan, A. Z., Pisella, L., Delporte, L., Rode, G., & Rossetti, Y. (2013, July 25). Testing for Optic Ataxia in a Blind Field. *Frontiers in Human Neuroscience*, *7*(Article 399). doi:10.3389/fnhum.2013.00399
- Koch, J. M., Datta, G., Makhdoom, S., & Grossberg, G. T. (2005). Unmet visual needs of Alzheimer's disease patients in long-term care facilities. *Journal of American Medical Directors Association*, *6*(4), 233 - 237.
- Krueger, R. A., & Casey, M. A. (2000). *Focus Groups: A Practical Guide for Applied Research* (3rd ed.). London: Sage Publications.

- Kumar, C. D., & Seet, E. (2016). Cataract surgery in dementia patients—
time to reconsider anaesthetic options. *British Journal of
Anaesthesia* 117 (4): 421–5 (2016), 117(4), 421-425.
doi:10.1093/bja/aew301
- Kumar, R. (1996). *Research Methodology: A Step-by-Step guide for
Beginners*. London: Sage Publications.
- Landis, T., Cummings, J. L., Benson, D. F., & Palmer, E. P. (1986, February).
Loss of topographic familiarity. An environmental agnosia. *Archives
of Neurology* 1986 Feb;43(2):132-6, 43(2), 132-136. Retrieved
March 2, 2018
- Larson, E. B., Shadlen, M.-F., Wang, L., McCormick, W. C., & al, e. (2004).
Survival after Initial Diagnosis of Alzheimer Disease. *Annals of
Internal Medicine*; Apr 6, 2004; 140, 7, 140(7), 501-509. Retrieved
February 26, 2018, from [https://search-proquest-
com.libproxy.ucl.ac.uk/docview/222222827/fulltextPDF/E633BE3F
09BD42E3PQ/1?accountid=14511](https://search-proquest-com.libproxy.ucl.ac.uk/docview/222222827/fulltextPDF/E633BE3F09BD42E3PQ/1?accountid=14511)
- Lawrence, V., Murray, J., Ffytches, D., & Banerjee, S. (2008). *The
experiences and needs of people with dementia and serious visual
impairment: a qualitative study*. London: Thomas Pocklington
Trust.
- Lawton, M. P. (2001). The physical environment of the person with
Alzheimer's disease. *Aging and Mental Health*, 17(4), 56 - 64.
- Lecouturier, J., Bamford, C., Hughes, J. C., Foy, R., Johnston, M., & Eccles,
M. P. (2008). Appropriate disclosure of a diagnosis of dementia
identifying the key behaviours of 'best practice'. *BMC Health
Services Research*. Retrieved September 28th, 2012, from BMC
Health Services Research: [http://www.biomedcentral.com/1472-
6963/8/95](http://www.biomedcentral.com/1472-6963/8/95)

- Lee, A. G., & Martin, C. O. (2004, February). Neuro-ophthalmic findings in the visual variant of Alzheimer's disease. *Ophthalmology*, *111*(2), 376-380.
- Leibowitz, H. M., Krueger, D. E., Maunder, L. R., & al, e. (1980, May/June). The Framingham Eye Study Monograph: An ophthalmological and epidemiological study of cataract, glaucoma, diabetic retinopathy, macular degeneration and visual acuity in a general population of 2631 adults, 1973 - 1975. *Survey of Ophthalmology*, *24*(Supplement), 335 - 610.
- Levine, D. N., Lee, J. M., & Fisher, C. M. (1993). The visual variant of Alzheimer's disease: a clinicopathologic case study. *Neurology*, *43*, 305-313.
- Liu, Y., Zhang, S., Tomata, Y., Nurrika, D., Sugawara, Y., & Tsuji, I. (2020). The impact of risk factors for dementia in China. *Age and Ageing*, *49*(5), 850-855. doi:10.1093/ageing/afaa048
- Livingston, G., Sommerlad, A., Orgeta, V., Costafreda, S. G., Huntley, J., Ames, D., . . . Mukadam, N. (2017). Dementia prevention, intervention, and care. *Lancet*(390), 2673-734. doi:10.1016/S0140-6736(17)31363-6
- Looney, T. (2016). *NVIVO in 7 steps*. London: ST Publishing House.
- Macular Society. (2017, May 31). *Global summit to address epidemic of age-related macular degeneration*. Retrieved February 28, 2018, from Macular Society:
<https://www.macularsociety.org/news/global-summit-address-epidemic-age-related-macular-degeneration>
- Mahoney, D. F., Jones, R. N., Coon, D. W., Mendelsohn, A. B., Gitlin, L. N., & Ory, M. (2003). The Caregiver Vigilance Scale: Application and validation in the Resources for Enhancing Alzheimer's Caregiver

Health (REACH) project. *American Journal of Alzheimers disease and other dementias*, 18, 39 - 48.

Manford, M., & Andermann, F. (1998). Complex Visual Hallucinations. *Brain*, 121, 1819-1840. Retrieved March 2, 2018

Manthorpe, J. (2001). Ethical ideals and practice. In C. Cantley (Ed.), *A Handbook in Dementia Care* (pp. 186-198). Maidenhead: Open University Press.

Marriam, S. B., & Tisdell, E. J. (2016). *Qualitative Research: A Guide to Design and Implementation* (4th ed.). San Francisco, CA: Jossey-Bass.

Marshall, M. (1998). Therapeutic Buildings for People with Dementia. In S. Judd, M. Marshall, & P. Phippen (Eds.), *Design for Dementia* (pp. 11 - 14). London: Journal of Dementia Care.

Maurer, K., Volk, S., & Gerbaldo, H. (1997). Auguste D and Alzheimer's disease. *Lancet*, 349(9064), 1546-1549.
doi:[https://doi.org/10.1016/S0140-6736\(96\)10203-8](https://doi.org/10.1016/S0140-6736(96)10203-8)

McCarthy, R. A., & Warrington, E. K. (1986). Visual associative agnosia: a clinico-anatomical study of a single case. *Journal of Neurology, Neurosurgery, and Psychiatry* 1986;49:1233-1240, 49, 1233-1240.
Retrieved March 1, 2018

McDonald, M., Hertz, R. P., Unger, A. N., & Lustik, M. B. (2009). Prevalence, Awareness, and Management of Hypertension, Dyslipidemia, and Diabetes Among United States Adults Aged 65 and Older. *J Gerontol A Biol Sci Med Sci*, 64A(2), 256 - 263.

McGowan, D. F. (1993). *Living in the Labyrinth*.

McIntyre, A., Harding, E., Yong, K. X., Sullivan, M. P., Gilhooly, M., Gilhooly, K., . . . Crutch, S. (2018). Health and social care practitioners' understanding of the problems of people with dementia-related

visual processing. *Health and Social Care in the Community*, 27,
982-990. doi:10.1111/hsc.12715

McMonagle, P., Deering, F., Berliner, Y., & Kertesz, A. (2006, February). The
cognitive profile of posterior cortical atrophy. *Neurology*, 66, 331-
338. Retrieved April 10th, 2017

McNair, D., Cunningham, C., Pollock, R., & McGuire, B. (2013). *Light and
lighting design for people with dementia*. Stirling: DSDC.

Meek, B. P., Shelton, P., & Marotta, J. J. (2013, June 13). Posterior Cortical
Atrophy: Visuomotor deficits in reaching and grasping. *Frontiers of
Human Neuroscience*, 7(Article 294), 1-16.
doi:10.3389/fnhum.2013.00294

Mendez, M. F. (2004). Posterior Cortical Atrophy: A Visual Variant of
Alzheimer's Disease. In A. Cronin-Golomb, & P. R. Hof (Eds.), *Vision
in Alzheimer's Disease* (Vol. 34, pp. 112-126). New York: Karger.

Mendez, M. F., Ghajaranian, N., & Perryman, K. M. (2002, June). Posterior
cortical atrophy: Clinical characteristics and differences compared
to Alzheimer's disease. *Dementia and Geriatric Cognitive Disorders*,
14(1), 33-40.

Mendez, M. F., Shapira, J. S., & Clark, D. G. (2007). Apperceptive Alexia in
posterior cortical atrophy. *Cortex*(43), 264-270. Retrieved July 2,
2016

Merriam, S. B., & Tisdell, E. J. (2016). *Qualitative Research: A Guide to
Design and Implementation*. San Francisco CA: John Wiley & Sons
Inc.

Meszaros, L. (2014, June 1st). *Is there a link between glaucoma and
dementia?* Retrieved April 7th, 2017, from Ophthalmology Times:
<http://ophthalmologytimes.modernmedicine.com/ophthalmologytimes/content/tags/2014-glaucoma-360/there-link-between-glaucoma-and-dementia?page=full>

- Metzler-Baddeley, C., Baddeley, R. J., Lovell, P. G., Laffan, A., & Jones, R. W. (2010). Visual Impairments in Dementia with Lewy Bodies and Posterior Cortical Atrophy. *Neuropsychology*, *24*(1), 35 - 48. doi: 10.1037/a0016834
- MHA. (2018). *MHA advises Emmerdale on dementia storyline*. Retrieved April 25, 2018, from MHA: https://www.mha.org.uk/news_archive/mha-advises-emmerdale-dementia-storyline/
- Midorikawa, A., Nakamura, K., Nagao, T., & Kawamura, M. (2008). Residual Perception of Moving Objects: Dissociation of Moving and Static Objects in a Case of Posterior Cortical Atrophy. *European Neurology*, *59*, 152-158. doi:10.1159/000114035
- Miles, M. B., & Huberman, A. M. (1994). *Qualitative Data Analysis*. Thousand Oaks, CA: Sage.
- Minassian, D., & Reidy, A. (2009). *Future Sight Loss UK (2): An epidemiological and economic model for sight loss in the decade 2010-2020: Full Report*. London: RNIB.
- Mitchell, P., Hayes, P., & Wang, J. J. (1997, Oct 20). Visual impairment in nursing home residents: the Blue Mountains Eye Study. *Medical Journal of Australia*, *167*(8), 416.
- Mizuno, M., Saror, G., Liccione, D., Battelli, L., & Campo, R. (1996). Progressive visual agnosia with posterior cortical atrophy. *Clinical Neurology and Neurosurgery*, *98*, 176-178. Retrieved March 1, 2018
- Moore, D. (2009). *Dementia: Guide to Dementia Care*. Brighton: Emerald Publishing.
- Morse, A. R. (2000). Vision and dementia. In B. Silverstone (Ed.), *The Lighthouse Handbook on Vision Impairment and Vision Rehabilitation* (Vol. 1). New York: Oxford University Press.

- Moustakas, C. (1994). *Phenomenological Research Methods*. London: Sage Publications Ltd.
- Munier, A., Gunning, T., Kenny, D., & O'Keefe, M. (1998). Causes of blindness in the adult population of the Republic of Ireland. *British Journal of Ophthalmology*, *82*(6), 630 - 633.
- Murgatroyd, C., & Prettyman, R. (2001). An investigation of visual hallucinosis and visual sensory status in dementia. *International Journal of Geriatric Psychiatry*, *16*, 709 - 713.
- Nagaratnam, N., Nagaratnam, K., Jolley, D., & Ting, A. (2001). Dementia following posterior cortical atrophy - a descriptive clinical case report. *Archives of Gerontology and Geriatrics*, *33*, 179 - 190.
- National Alliance for Caregiving. (2011). *What made you think Mom had Alzheimer's?* Maryland: National Alliance for Caregiving.
- National Audit Office. (2010). *Improving Dementia Services in England - an Interim Report*. London: The Stationery Office.
- National Collaborating Centre for Mental Health . (2007). *National Clinical Practice Guideline Number 42: NICE–SCIE Guideline on supporting people with dementia and their carers in health and social care*. London: The British Psychological Society and Gaskell.
- National Eye Institute (NEI). (2015, September). *Facts About Diabetic Eye Disease*. Retrieved April 7, 2017, from National Institutes of Health: <https://nei.nih.gov/health/diabetic/retinopathy>
- Neitzel, J., Ortner, M., Haupt, M., Redel, P., Grimmer, T., Yakushev, I., . . . Fink, K. (2016). Neuro-cognitive mechanisms of simultanagnosia in patients with posterior cortical atrophy. *Brain*, *139*, 3267-3280. doi:10.1093/brain/aww235
- Newbronner, L., Chamberlain, R., Borthwick, R., Baxter, M., & Glendinning, C. (2013). *A Road Less Rocky: Supporting Carers of People with Dementia*. London: Carers Trust.

NHS. (2018, July). *Eye health tips for older people*. Retrieved from NHS UK:
<https://www.nhs.uk/live-well/healthy-body/eye-health-tips-for-older-people/>

NHS Choices. (2015, June 22). *Dementia Guide: Managing legal affairs for someone with dementia*. Retrieved May 23, 2018, from NHS Choices: <https://www.nhs.uk/conditions/dementia/legal-issues/>

NICE. (2016, September). *Dementia: supporting people with dementia and their carers in health and social care*. Retrieved April 25, 2018, from NICE:
<https://www.nice.org.uk/guidance/cg42/ifp/chapter/What-is-dementia>

NICE. (2019). *Dementia: Quality Standard*. London: NICE.

NINDS. (2017, May 24). *Prosopagnosia Information Page*. Retrieved March 3, 2018, from National Institute of Neurological Disorders and Stroke: <https://www.ninds.nih.gov>

Norton, S., Matthews, F. E., & Brayne, C. (2013). A commentary on studies presenting projections of the future prevalence of dementia. *BMC Public Health, 13*, 1.

Nowell, Z. K., Thornton, A., & Simpson, J. (2011). The subjective experiences of personhood in dementia care settings. *Dementia, 12*(4), 394 - 409. doi:10.1177/1471301211430648

O'Connor, D., Phinney, A., Smith, A., Small, J., Purves, B., Perry, J., . . . Beattie, L. (2007). Personhood in dementia care: Developing a research agenda for broadening the vision. *Dementia*(6), 121-142.

Ohno-Matsui, K. (2011). Parallel findings in age-related macular degeneration and Alzheimer's disease. *Progress in Retinal and Eye Research, 30*, 217 - 238.

Ortiz-Figueroa, Y. B., Perez, A. G., Perez, R. C., Graue-Hernandez, E. O., Arrieta, J., & Jimenez-Corona, A. (2016, September). Improvement

in the quality of life after cataract surgery in patients older than 50 years of age. *Investigative Ophthalmology & Visual Science*, 57, 6233.

Owen, C. G., Jarrar, Z., Wormald, R., Cook, D. G., Fletcher, A. E., & Rudnicka, A. (2012). The estimated prevalence and incidence of late stage age related macular degeneration in the UK. *British Journal of Ophthalmology*, 96, 752-756. doi:10.1136/bjophthalmol-2011-3011

Pankow, L. J., Pryor, J. L., & Luchins, D. J. (2009). 'Glaucoma Screening of Patients With and Without Dementia', 6: 1, 29 — 39. *Journal of Evidence-Based Social Work*, 6, 29-39.

Parkin, E., & Baker, C. (2016). *Dementia: policy, services and statistics: Briefing Paper No 7007*. London: House of Commons Library.

Parkinson, M., Carr, S. M., & Rushmer, R. (2016, September 27). Investigating what works to support family carers of people with dementia: a rapid realist review. *Journal of Public Health*, 39(4), e290–e301. doi:10.1093/pubmed/fdw100 |

Perrin, T. (1997). Occupational need in severe dementia: a descriptive study. *Journal of Advanced Nursing*, 25, 934-941.

Perrin, T., May, H., & Anderson, E. (2008). *Wellbeing in Dementia: An Occupational Approach for Therapists and Carers*. Edinburgh: Church Livingstone Elsevier.

Peterson, B. (2004). *Voices of Alzheimer's: courage, humor, hope and love in the face of dementia*. Cambridge, MA: De Capo Books.

Pool, J. (2010). *Alzheimer's Society guide to the dementia care environment*. London: Alzheimer's Society.

Porter, D. (2017, March 1). *What are Drusen?* Retrieved from American Academy of Ophthalmology: <https://www.aao.org/eye-health/diseases/what-are-drusen>

- Power, G. A. (2014). *Dementia Beyond Disease: Enhancing Well-being*. Baltimore: Health Professions Press.
- Pratchett, T. (2015). *Shaking Hands with Death*. London: Transworld Publishers.
- Pratt, R., & Wilkinson, H. (2001). *Tell me the Truth: The effect of being told the diagnosis of dementia from the perspective of the person with dementia*. London: The Mental Health Foundation.
- QSR International. (2017). NVIVO Pro: Version 11. London, United Kingdom. Retrieved from <http://www.qsrinternational.com/nvivo/what-is-nvivo>
- Quadi, N., Alipour, S., & Beattie, B. L. (2007). Pathophysiology of Alzheimer's disease. In H. H. Feldman (Ed.), *Atlas of Alzheimer's Disease* (pp. 59-69). Andover: Informa UK Ltd.
- Rahman, S. (2014). *Living Well with Dementia: The importance of the Person and the Environment for Wellbeing*. London: Radcliffe Publishing.
- Raina, P., Wong, M., & Massfeller, H. (2004). The relationship between sensory impairment and functional independence among elderly. *BMC Geriatrics*.
- Rait, G., Fletcher, A., Smeeth, L., Brayne, C., Stirling, S., Nunes, M., . . . Tullock, A. J. (2005). Prevalence of cognitive impairment: results from the MRC trial of assessment and management of older people in the community. *Age and Ageing*, 34(3), 242 - 248.
- Ramachandran, V. S., Altschuler, E. L., & Hillyer, S. (1997, May 22). Mirror agnosia. *Proceedings: Biological Sciences*, 264(1382), 645-647. Retrieved March 2, 2018, from <http://www.jstor.org/stable/51082>
- Rare Dementias Support (UCL). (2021, January 21). *Posterior Cortical Atrophy*. Retrieved from Rare Dementias Support: <https://www.raredementiasupport.org/posterior-cortical-atrophy/>

- Ray, S., & Davidson, S. (2014). *Dementia and Cognitive Decline: A Review of the Evidence*. London: AgeUK Research.
- Reidy, A., Minnassian, D. C., Valfidis, G., Joseph, J., Farrow, S., Wu, J., . . . Connolly, A. (1998). Prevalence of serious eye disease and visual impairment in a north London population: Population based cross sectional study. *British Medical Journal*(316), 1643-1646.
- Reisberg, B., Ferris, S. H., de Leon, M. J., & Crook, T. (1982). The Global Deterioration Scale for assessment of primary degenerative dementia. *American Journal of Psychiatry*(139), 1136-9.
- Renner, J. A., Burns, J. M., Hou, C. E., McKeel Jr, D. W., Storandt, M., & Morris, J. C. (2004 Oct 12;63(7):1175-80.f.). Progressive posterior cortical dysfunction: a clinicopathologic series. *Neurology*, 63(7), 1175-80. doi:10.1212/01.wnl.0000140290.80962
- Ressiniotis, T., Griffiths, P. G., Birch, M., Keers, S. M., & Chinnery, P. F. (2004). Apolipoprotein E promoter polymorphisms do not have a major influence on the risk of developing primary open angle glaucoma. *Molecular Vision*, 10, 805 - 807.
- RNIB. (2014, September 5). *Time for your exercise*. Retrieved May 23, 2018, from RNIB: <https://www.rnib.org.uk/time-your-exercise>
- RNIB. (2016). *Age-related macular degeneration*. London: RNIB.
- RNIB. (2020). *Cataracts and dementia*. Retrieved February 24, 2021, from RNIB: Visit www.vision2020uk.org.uk
- RNIB. (2021, April 20). *Technology*. Retrieved from RNIB: <https://shop.rnib.org.uk/technology>
- Roberts, K. L., & Allen, H. A. (2016, March 1). Perception and Cognition in the Ageing Brain: A Brief Review of the Short- and Long-Term Links between Perceptual and Cognitive Decline. *Frontiers in Aging Neuroscience*. Retrieved April 7th, 2017, from

<http://journal.frontiersin.org/article/10.3389/fnagi.2016.00039/ful>

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- Roco, M., Gleichgerrcht, E., Torralva, T., & Manes, F. (2010). Cognitive rehabilitation in posterior cortical atrophy. *Neuropsychological Rehabilitation, 20*(4), 528-540. doi:10.1080/09602011003597408
- Rogelet, P., Delafosse, A., & Destee, A. (1996). Posterior cortical atrophy: unusual feature of Alzheimer's disease. *Neurocase, 2*, 495-501.
- Rose, H. (1996). *Show Me The Way To Go Home*. Forest Knolls: Elder Books.
- Ryan, N. S., Shakespeare, T. J., Lehmann, M., Keihaninejad, S., Nicholas, J. M., Leung, K. K., . . . Crutch, S. J. (2014). Motor features n posterior cortical atrophy and their imaging correlates. *Neurobiology of Aging, 35*(12), 1-13.
- Sacks, O. (1986). *The Man who Mistook his Wife for a Hat*. London: Picador.
- Sacks, O. (2010). *The Mind's Eye*. London: Picador.
- Scafer, D. (2002). 'That Senescence Itself Is an Illness': A Transitional Medical Concept of Age and Ageing in the Eighteenth Century. *Medical History, 46*(4), 525-548.
- Schott, J. M., & Crutch, S. J. (2019, February). Posterior Cortical Atrophy. *CONTINUUM, 52* - 75. Retrieved January 3rd, 2020
- Schumacher, L. A. (2010). *The caregiver's journey: a phenomenological study of the lived experience of leisure for caregivers in the sandwich generation who care for a parent with dementia.* " MA (Master of Arts) thesis,. Iowa: University of Iowa. doi:10.17077/etd.g638o927
- SCIE. (2015, May). *Dementia*. Retrieved May 16, 2018, from Social Care Institute for Excellence: <https://www.scie.org.uk/dementia/about/>
- SCIE. (2015, May). *Dementia: Advance care planning in dementia*. Retrieved April 23, 2018, from Social Care Institute for Excellence (SCIE):

<https://www.scie.org.uk/dementia/supporting-people-with-dementia/decisions/advance-care-planning.asp>

Scrutton, J., & Brancati, C. U. (2016). *Dementia and comorbidities*. London: ILC-UK.

Shakespeare, T. J., Yong, K. X., Foxe, D., Hodges, J., & Crutch, S. J. (2015). Pronounced Impairment of Everyday Skills and Self-Care in Posterior Cortical Atrophy. *Journal of Alzheimer's Disease*, *43*, 381-384. doi:DOI 10.3233/JAD-141071

Shakespeare, T. J., Yong, K. X., Frost, C., Kim, L. G., Warrington, E. K., & Crutch, S. J. (2013, October 2). Scene Perception in Posterior Cortical Atrophy: categorization, description and fixation patterns. *Frontiers in Human Neuroscience*, *7*(Article 621). doi:10.3389/fnhum.2013.00621

Sinclair, A., Hinds, A., & Sanders, R. (2004). Ten years of glaucoma blindness in Fife 1990 - 99 and the implications for ophthalmology, optometry and rehabilitation services. *Ophthalmic and Physiological Optics*, *24*, 313 - 318.

Smith, J. A., Flowers, P., & Michael, L. (2009). *Interpretative Phenomenological Analysis: Theory, Method and Research*. London: Sage Publications Ltd.

Smith, J., & Osborn, M. (2007). Pain as an assault on the self: An interpretative phenomenological analysis of the psychological impact of chronic benign low back pain. *Psychology and Health*, *22*(5), 517-534. doi:10.1080/14768320600941756

Snowdon, J. S., Stopford, C. I., Julien, C. L., Thompson, J. C., Davidson, Y., Gibbons, L., . . . Mann, D. M. (2007). Cognitive Phenotypes in Alzheimer's Disease and Genetic Risk. *Cortex*, *43*(7), 835-845. doi:10.1016/S0010-9452(08)70683-X

- Stake, R. E. (1995). *The Art of Case Study Research*. London: Sage Publications.
- Stanley, D., & Cantley, C. (2001). Assessment, care planning and care management. In C. Cantley (Ed.), *A Handbook of Dementia Care* (pp. 109-122). Maidenhead: Open University.
- Stephen, B., & Brayne, C. (2008). Prevalence and projections of dementia. In M. Downs, & B. Bowers (Eds.), *Excellence in Dementia Care: Research into Practice* (pp. 9-34). Maidenhead: Open University Press.
- Stokes, G. (2010). *And Still the Music Plays: Stories of People with Dementia* (2nd Edition ed.). London: Hawker Publications.
- Strategy and Projects Team, Care and Transformation. (2018). *Carers Action Plan 2018-2020: Supporting Carers Today*. London: Department of Health and Social Care. Retrieved April 1, 2021
- Suarez-Gonzalez, A., Crutch, S. J., Franco-Macias, E., & Gil-Neciga, E. (2016). Neuropsychiatric Symptoms in Posterior Cortical Atrophy and Alzheimer Disease. *Journal of Geriatric Psychiatry, 29*, 65-71. doi:10.1177/0891988715606229
- Suarez-Gonzalez, A., Ocal, D., Pavisic, I., Peacock, A., Naessan, M., Ahmed, S., . . . Crutch, S. (2019). ReadClear: An Assistive Reading Tool for People Living with Posterior Cortical Atrophy'. *Journal of Alzheimer's Disease, 71*(4), 1285-1295. doi:10.3233/JAD-190335
- Talan, J. (2011, May 5). New Data: Posterior Cortical Atrophy Shares Similarities with Alzheimer Disease. *Neurology Today, 37*. Retrieved April 10th, 2017
- Talan, J. (2017, April 6). International Consensus on the First Diagnostic Guidelines for Posterior Cortical Atrophy. *Neurology Today, 17*(7), 1-19-20.

- Tang-Wai, D. F., & Graff-Radford, N. R. (2011). Looking into posterior cortical atrophy: Providing insight into Alzheimer disease. *Neurology, 76*, 1778-1779. Retrieved July 4, 2016
- Tang-Wai, D. F., Graff-Radford, N. R., Boeve, B. F., Dickson, D. W., Parisi, J. E., Crook, R., . . . Petersen, J. C. (2004). Clinical, genetic, and neuropathologic characteristics of posterior cortical atrophy. *Neurology, 63*(7), 1168-1174.
doi:10.1212/01.WNL.0000140289.18472.15
- Tang-wai, D. F., Josephs, K. A., Boeve, B. F., & et al. (2003). Pathologically confirmed corticobasal degeneration presenting with visuospatial dysfunction. *Neurology, 61*(8), 1134-1135.
doi:10.1136/jnnp.61.4.388
- Tang-Wai, D., & Mapstone, M. (2006). What are we seeing? Is posterior cortical atrophy just Alzheimer disease? *NEUROLOGY*(66), 300-301.
Retrieved July 2, 2016
- Taylor, B. (1993). Phenomenology: one way to understand nursing practice. *International Journal of Nursing Studies, 30*(2), 171-179.
doi:10.1016/0020-7489(93)90066-4
- The Lewy Body Society. (2015, June). *Polka Dot-ty days*. Retrieved May 1st, 2015, from lewybody.org:
<http://www.lewybody.org/download/dotty-days-lewy-body-awareness/?wpdmdl=6685>
- Thibaud, M., Bloch, F., Tournoux-Facon, C., Breque, Sophie, A., Dugue, B., & Kemoun, G. (2012, April). Impact of physical activity and sedentary behaviour on fall risks in older people: a systematic review and meta-analysis of observational studies. *European Review of Aging and Physical Activity, 5*(1), 5-15. doi:10.1007/s11556-011-0081-1
- Thomas, G. (2011). *How to do Your Case Study: A Guide for Students and Researchers*. London: Sage Publications Ltd.

- Thomas, G. (2016). *How to do your Case Study* (2nd ed.). London: Sage.
- Tideiksaar, R. (2002/3, Winter). Tideiksaar, R. (2002/3, Winter). Sensory Impairment and Fall Risk. *Generations*, 22 - 27. *Generations*, 22-27.
- Tom, S. E., Hubbard, R. A., Crane, P. K., Haneuse, S. J., Bowen, J., McCormick, W. C., . . . Larson, E. B. (2015). Characterization of Dementia and Alzheimer' s Disease in an Older Population: Updated Incidence and Life Expectancy With and Without Dementia. *American Journal of Public Health*(105), 408-413. doi:10.2105/AJPH.2014.301935
- Tondo, G., De Marchi, F., Terazzi, E., Sacchetti, M., & Cantello, R. (2017). A Case of Posterior Cortical Atrophy with Complex Set of Symptoms and Rapid Course. *JOURNAL OF NEUROLOGY AND NEUROSCIENCE*, 8(3), 196. doi:10.21767/2171-6625.1000196
- Trigg, R., & Jones, R. (2005). *Dementia and Blindness*. London: Thomas Pocklington Trust.
- Tsai, D. C., Chen, S. J., Huang, C. C., Yaun, M. K., & Leu, H. B. (2015). Age-Related Macular Degeneration and Risk of Degenerative Dementia among the Elderly in Taiwan: A Population-Based Cohort Study. *Ophthalmology*, 122(11), 2327 - 2335. doi:10.1016/j.optha.2015.07.033
- Tsai, P. H., & Mendez, M. F. (2009, September 1). AKINETOPSIA IN THE POSTERIOR CORTICAL VARIANT OF ALZHEIMER DISEASE. 73(9), 731-732. doi:https://doi.org/10.1212/WNL.0b013e3181b59c07
- UCAF Memory and Aging Center. (2014, July 1). *Posterior Cortical Atrophy*. Retrieved April 30th, 2017, from UCSF Memory and Aging Center: <http://memory.ucsf.edu/education/diseases/pca>
- UK Office for National Statistics. (2016). *Mid-2015 Population Estimates*. London: Office for National Statistics.

- Utton, D. (2007). *Designing Homes for People with Dementia*. London: The Journal of Dementia Care.
- Valenti, D. (2004). The Anterior visual System and Circadian Function with Reference to Alzheimer's Disease. In *Vision in Alzheimer's Disease* (pp. 1-29). New York: Karger.
- van Manen, M. (1997). *Researching Lived Experience: Human Science for an Action Sensitive Pedagogy*. London: Routledge.
- Van Manen, M. (2016). *Phenomenology of Practice: Meaning-Giving Methods in Phenomenological Research and Writing*. Abingdon: Routledge.
- Van Manen, M. (2016). *Researching Lived Experience: Human Science for an Action Sensitive Pedagogy* (2nd ed.). Abingdon: Routledge.
- Vann Jones, S. A., & O'Brien, J. T. (2014, March). The prevalence and incidence of dementia with Lewy bodies: a systematic review of population and clinical studies. *Psychological Medicine*, 673-683. Retrieved March 30, 2017, from <https://doi.org/10.1017/S0033291713000494>
- Vickers, J. C., Craig, J. E., Stankovich, J., McCormack, G. H., West, A. K., Dickinson, J. L., . . . Mackey, D. A. (2002). The apolipoprotein $\epsilon 4$ gene is associated with elevated risk of normal tension glaucoma. *Molecular Vision*, 8, 389 - 393.
- Victor, C. R., Rippon, I., Nelis, S. M., Martyr, A., Litherland, R., Pickett, J., . . . Clare, L. (2020). Prevalence and determinants of loneliness in people living with dementia: Findings from the IDEAL programme. *International Journal of Geriatric Psychiatry*, 35, 851-858. doi:10.1002/gps.5305
- Victoroff, J., Webster Ross, G., Benson, D. F., Verity, A., & Vinters, H. V. (1994). Posterior Cortical Atrophy. *Archives of Neurology*, 51(3), 269-274. doi:10.1001/archneur.1994.00540150063018

- Videaud, H., Torny, F., Cartz-Piver, L., Deschamps-Vergara, N., & Couratier, P. (2012). [Impact of drug-free care in posterior cortical atrophy: Preliminary experience with a psycho-educative program. *Rev Neurology (Paris)*, *168*, 861-867. doi:10.1016/j.neurol.2011.10.013
- Wahl, H. W., & Heyl, V. (2003). Connections between vision, hearing and cognitive function in old age. *Generations, Spring*, 39 - 45.
- Ward, M. E., & Petersen, R. C. (2013, August). Glaucoma and Dementia: More Than Meets the Eye? *Annals of Neurology*, *74*(2), 155-157. doi:10.1002/ana.23949
- Warren, J. D., Rohrer, J. D., & Rossor, M. N. (2013, August 6). Frontotemporal dementia. *BMJ*, *347*. Retrieved May 1st, 2017, from <http://www.bmj.com/content/bmj/347/bmj.f4827.full.pdf>
- Warrington, E. K., & James, M. (1988). Visual apperceptive agnosia: a clinico-anatomical study of three cases. *Cortex*, 13-32. Retrieved March 2, 2018
- Weill-Chounlamountry, A., Alves, J., & Pradat-Diehl, P. (2016). Non-pharmacological intervention for posterior cortical atrophy. *World Journal of Clinical Cases*, *4*(8), 195-201. doi:10.12998/wjcc.v4.i8.195
- Weill-Chounlamountry, A., Poncet, F., Crop, S., Hesly, N., Mouton, A., Samri, D., . . . Pradat-Diehl, P. (2012). Physical medicine and rehabilitation multidisciplinary approach in a case of Posterior Cortical Atrophy. *Annals of Physical and Rehabilitation Medicine*, *55*, 443-449. doi:10.12998/wjcc.v4.i8.195
- Wertz, F. J., Charmaz, K., McMullen, L. M., Josselson, R., Anderson, R., & McSpadden, E. (2011). *Five Ways of Doing Qualitative Analysis*. New York: The Guildford Press.
- Whitman, L. (2016). *People with Dementia Speak Out*. London: Jessica Kingsley Publishers.

- Whitwell, J. L., Jack Jr, C. R., Kantarci, K., Weigand, S. D., Boeve, B. F., & et al. (2007, July). Imaging correlates of posterior cortical atrophy. *Neurobiology of Aging*, 28(7), 1051-1061.
doi:10.1016/neurobiolaging.2006.05.026
- Wilkinson, H. (2002). Including people with dementia in research: methods and motivations. In H. Wilkinson (Ed.), *The perspectives of people with dementia: research methods and motivations* (p. 9). London: Jessica Kingsley Publishers.
- Willis, J. R., Vitale, S. E., Agrawal, Y., & Ramulu, P. Y. (2013, June 6). Visual Impairment, Uncorrected Refractive Error, and Objectively Measured Balance in the United States. *JAMA Ophthalmology*, 131(8), 1049-1056. doi:10.1001/jamaophthalmol.2013.316
- Wostyn, P., Audenaert, K., & De Deyn, P. P. (2009). Alzheimer's disease and glaucoma: Is there a causal relationship? *British Journal of Ophthalmology*, 93, 1557 - 1559.
- Yin, R. K. (2009). *Case Study Research: Design and Methods* (4th ed.). London: Sage Publications.
- Yong, K. X., Rajdev, K., Shakespeare, T. J., Leff, A. P., & Crutch, S. J. (2015). Facilitating text reading in posterior cortical atrophy. *Neurology*, 85(4), 339-348.
doi:https://dx.doi.org/10.1212%2FWNL.0000000000001782
- Zakzanis, K. K., & Boulos, M. I. (2001). Posterior Cortical Atrophy. *The Neurologist*(7), 341-349. Retrieved July 1, 2016
- Zetterberg, M., Landgren, S., Andersson, M. E., Palmer, M. S., Gustafson, D. R., Skoog, I., . . . Zetterberg, H. (2008). Association of Complement Factor H Y402H Gene Polymorphism with Alzheimer's Disease. *American Journal of Medical Genetics Part B (Neuropsychiatric Genetics)*, 147B, 720 - 726.

Appendices

Appendix One: Stages of Dementia described by the Global Deterioration Scale

Stages of Dementia described by the Global Deterioration Scale	
Global Deterioration Scale Stages	General Features
Stage 1 (No cognitive decline)	No subjective symptoms or clinical evidence of memory deficit
Stage 2 (Very mild cognitive decline)	Subjective symptoms of memory deficit, such as forgetting the location of familiar objects or previously well-known names.
Stage 3 (Mild cognitive decline)	Early, clear-cut memory deficits, such as getting lost, poor work performance, problems with word or name finding, misplacing objects of value, or poor retention of written material
Stage 4 (Moderate cognitive decline – <i>early dementia</i>)	Clear-cut memory deficits, such as poor knowledge of current events, difficulty remembering details of personal history, impaired concentration doing serial subtractions, and inability to handle finances or travel to family locations. Average duration 2 years.
Stage 5 (Moderately severe cognitive decline - <i>middle dementia</i>)	Can no longer function without some assistance. Unable to recall major aspects of current life (e.g., address, telephone number, names of family members). May need assistance dressing, but still independent in eating and toileting. Average duration 1.5 years
Stage 6 (Severe cognitive decline – <i>middle dementia</i>)	Occasionally forgets name of primary caregiver (e.g., spouse). Largely unaware of recent personal events. Substantial assistance required for activities of daily living and travel to familiar locations. Often unaware of surroundings (e.g. year or season). Can still recall his or her own name and distinguish a familiar face. Average duration 2.5 years
Stage 7 (Very severe cognitive decline – <i>Late dementia</i>)	Unable to recognise familiar faces. Verbal ability limited to less than 5 words. Incontinent of urine and stool. Total functional dependence, and unable to walk. Average duration 2.5 years
(Reisberg, Ferris, de Leon, & Crook, 1982)	

Appendix Two: Stages of Posterior Cortical Atrophy

Stages of Posterior Cortical Atrophy	
<p>Aims</p> <p>When given a diagnosis of PCA, many people naturally ask what will happen next, how long will they be able continue with a particular activity, or when and what care will they need. Such questions are often met with the response ‘we don’t know’ or ‘it’s different for each person’. In many ways these responses are reasonable, as the experience of PCA can indeed vary greatly from one person to the next. Symptoms may vary from individual to individual owing to a variety of factors including age, health and the disease underlying the PCA syndrome. Not everyone will experience the same symptoms or progress at the same rate. But whilst true, these responses are not terribly helpful.</p> <p>This document attempts to provide a general framework for describing how abilities may change during the course of PCA. It is hoped that terms such as the ‘stages’ described may provide a common language which helps when discussing which treatments, therapies, support or decisions are appropriate.</p>	
Global Deterioration Scale Stages	General Features
<p>Stage 1 <i>(No cognitive decline)</i></p>	<p>The person does not experience any visual problems, memory or other cognitive problems. An interview with a medical professional does not show any evidence of symptoms of vision loss or dementia.</p>
<p>Stage 2 <i>(Very mild cognitive decline) (may include problems only recognized in hindsight)</i></p>	<p>The person experiences subtle difficulties with some complex visual tasks e.g.</p> <ul style="list-style-type: none"> - a loss of confidence in their driving ability owing to finding it harder to judge distances - problems using a computer such as being slower to type or enter information - difficulties with reading, writing, spelling and/or arithmetic - difficulty recalling and writing down telephone numbers - difficulty assembling novel devices/objects <p>The person may experience vertigo or other balance problems (although some aspects of these may develop later or not at all), they may experience difficulties finding the right words when speaking or odd visual sensations (e.g. occasional changes or washes of colour in the centre or periphery of their vision).</p> <p>These problems may be intermittent, difficult to describe and not noticed by friends, family or co-workers. Insight into, but uncertainty as to the cause of these experiences may contribute to anxiety in some individuals. This stage may reflect the earliest</p>

	<p>signs of PCA, especially where eye tests reveal no indication of ophthalmological problems.</p>
<p>Stage 3 <i>(Mild cognitive decline)</i> <i>(Early-stage Posterior Cortical Atrophy can be diagnosed in some, but not all, individuals with these symptoms)</i></p>	<p>Visual difficulties are acknowledged both by the person in question and their friends, family and co-workers, and the person is likely to decide to stop driving (although some people may have already stopped by this stage). Healthcare professionals familiar with young onset and atypical dementias may recognize a fundamental difficulty in perceiving what and where things are leading to characteristic problems including:</p> <ul style="list-style-type: none"> - Difficulty seeing things which are ‘right under your nose’ or only seeing part of something (e.g. not noticing food on the edge of a plate) - General ‘clumsiness’ (in people who were not previously clumsy), such as knocking things over (inaccurate reaching out), putting a glass down sideways, etc. - Difficulty finding things in a handbag, cupboard, etc. - Difficulties dressing and performing other activities (e.g. DIY, cutting a piece of toast in half) that require spatial judgement (e.g. using stairs) and (e.g. left-right) coordination - Inability to tell the time from a clock or watch (especially analogue) - Getting ‘lost’ on the page when reading - Problems with writing, spelling and dealing with numbers - Paying for items with a note or letting vendors select coins for them (to avoid having to handle change) - Mild memory problems for recent events, though less pronounced than visual problems – these could occur for different reasons, e.g. able to remember many events very well but difficulty for example learning new tasks - Mild difficulty retrieving the correct word in conversation <p>These difficulties may be accompanied by anxiety; this may partly reflect awareness of the difficulties, but equally anxiety and feeling flustered can exacerbate the visual and other problems. People at this stage may also experience subtle sensory changes and disturbances (e.g. feeling cold).</p>
<p>Stage 4 <i>(Moderate cognitive decline – early dementia)</i> <i>(Mild or early-stage Posterior Cortical Atrophy)</i></p>	<p>At this point, individuals with PCA may require significant help with everyday activities such as cooking, dressing and using appliances. The person retains a clear sense of purpose regarding activities and plans but attempts to achieve these goals are frustrated by frequent, small visual problems and have significant implications for independence and the care/support required e.g.</p> <ul style="list-style-type: none"> - Inability to read labels on food packets - Problems discriminating coins

	<p>- Inability to tell the time from an analogue or even digital watch</p> <p>There may be significant contrasts between what people can and cannot do e.g.</p> <ul style="list-style-type: none"> - Dissociation in ability to walk, sit down and stand up (e.g. might be able to walk but not sit down or unable to stand up and walk). - Being able to see some things but not others (e.g. following moving targets better than localizing static objects; can't read but can still follow or at least get pleasure from TV or the cinema) <p>There may be changes in the ability to navigate around unfamiliar and familiar environments: Someone with PCA might be aware that they are at home but have difficulty navigating from one room to the next, or locating the bathroom; some may find a loss in their mental map resulting in problems navigating even familiar environments, (e.g. the home) or problems locating rooms (especially in low light). This might result in getting lost within the home. Problems outside the house might also occur.</p> <p>Severe visual and other problems may also result in:</p> <ul style="list-style-type: none"> - Inability to engage in a number of pastimes such as reading - An inability to identify even very familiar faces, with recognition of identity dependent upon hearing someone's voice - Resorting to eating with one's hands, or being fed, owing to difficulty manipulating cutlery and seeing the location of food on a plate <p>In addition, increasing word finding difficulties may mean that more time is required to express oneself, leading to reduced participation in group conversations.</p>
<p>Stage 5 <i>(Moderately severe cognitive decline - middle dementia)</i> <i>(Moderate or mid-stage Posterior Cortical Atrophy)</i></p>	<p>Vision deteriorates further to the extent that the world is viewed as individual pieces of a puzzle or as if through a fractured mirror, with occasional small details noticed but difficulty appreciating whole scenes or the relationship between different objects.</p> <p>Individuals with PCA may now:</p> <ul style="list-style-type: none"> - Require help with most or all everyday tasks, including toileting and personal care owing to problems undoing zips/buttons, positioning the body relative to furniture and seeing and feeling the position of clothing, etc. - Experience spatial problems not limited to vision, with spatial commands and actions (e.g. 'turn round', 'shuffle forwards') difficult to comprehend and execute

	<ul style="list-style-type: none"> - Require support and/or supervision when walking, for example because of difficulty detecting the edge of pavements, paths and steps etc., and/or because of a tendency to shuffle, stoop or lean <p>At this stage, many individuals are registered as blind, meaning they require support in all visually guided activities. Episodic memory and word finding difficulties may also now be prominent, though many individuals retain considerable awareness of their situation. Some people may have problems with multiple senses, e.g. problems with:</p> <ul style="list-style-type: none"> - Sensing the relative position of parts of the body (as pronounced as not knowing if one is ‘the right way up’) - Headache, increased sensitivity to pain and/or sensations in the scalp or other body parts, people with PCA may become more sensitive to the cold and may be prone to experiencing cold hands and feet - Sense of hearing may change: people with PCA may become more sensitive to certain sounds. - Feelings of imbalance or instability, particularly when walking - Little jerky movements in the fingers, arms or other parts of the body
<p>Stage 6 <i>(Severe cognitive decline – middle dementia)</i> <i>(Moderately severe or mid-stage Posterior Cortical Atrophy)</i></p>	<p>At this stage, cognitive changes are more global in nature, with multiple aspects of perception, memory, language, attention and decision-making abilities affected. Vision remains the most pronounced impairment, but in most individuals, there are widespread impairments that would be recognised as dementia. Individuals may:</p> <ul style="list-style-type: none"> - Be ‘functionally blind’, requiring support in all visually-guided activities - Show inconsistent recollection of recent events and plans - Show halting, non-fluent speech with difficulty retrieving many words and the production of sound-based speech errors - Experience changes in sleep patterns — sleeping during the day and becoming restless at night. - Have increasingly frequent trouble controlling their bladder or bowels – <p>Become quiet and withdrawn, sometimes sitting in a stooped over manner and appearing disengaged from the environment unless addressed directly</p> <ul style="list-style-type: none"> –Experience behaviour and personality changes, including obsessions, compulsions, suspiciousness and delusions (such as believing that their caregiver is an impostor). - Have difficulty communicating, as expressing and understanding verbal and visual stimuli becomes increasingly difficult. - Experience continued deterioration of sensory functions, e.g. partial or complete loss of response to touch

<p>Stage 7 (<i>Very severe cognitive decline – Late dementia</i>) (<i>Severe or late-stage Posterior Cortical Atrophy</i>)</p>	<p>In the late stages, the problems experienced by people with PCA may resemble typical AD more closely than at any stage earlier in the disease process. In the final stage of this disease, individuals lose the ability to respond to their environment, to carry on a conversation and, eventually, to control movement. They may still say words or phrases. There may be discrepancies e.g. inability to smile but continued ability to laugh. At this stage, individuals need help with much of their daily personal care, including eating or using the toilet. They may also lose the ability to smile, to sit without support and to hold their heads up. Reflexes become abnormal. Muscles grow rigid. Swallowing is impaired. Maintaining adequate nutrition, hydration and skin integrity can be challenging at this stage but problems (e.g. bedsores etc.) are not inevitable.</p>
<p>End of Life</p>	<p>Although Alzheimer’s disease and other degenerative diseases are life shortening illnesses, another condition or illness (such as pneumonia) may actually cause the person’s death. Pneumonia is listed as the cause of death in up to two thirds of people with dementia. The person’s ability to cope with infections and other physical problems will be impaired due to the progression of the disease. In some people no specific cause of death is found, other than Alzheimer’s disease. Depending on the circumstances, ‘Alzheimer’s disease’ or similar may be entered on the death certificate as the sole or main cause of death, or as a contributing factor.</p>
<p>(Crutch, Walton, Carton, & Shakespeare, 2017)</p>	

Appendix Three: Information sheet and consent form



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INFORMATION SHEET

Title of Project: *“A qualitative study exploring the specific everyday social and environmental challenges faced by community dwelling people living with posterior cortical atrophy (PCA)”*

Name of Researcher: Pam Turpin, University College London

You are being invited to take part in a research study. This research is part of a PhD being conducted by Pamela Turpin at University College London exploring what changes to social and physical environments could improve the daily lives of people with posterior cortical atrophy. This research is part of a larger study funded by Alzheimer’s Research UK/Economic and Social Research Council/National Institute for Health Research and is based at University College London (UCL) with involvement from UCL and Brunel University researchers. Before you decide it is important for you to understand why the research is being done and what it will involve. Please take time to read the following information carefully and discuss it with others if you wish. Ask us if you are not clear on any of the information. Take time to consider if you wish to take part.

What is the purpose of this study?

To examine the range, type and impact of visual problems experienced by individuals with Posterior Cortical Atrophy (PCA). Pam’s study is looking at the day to day challenges for people with PCA and what might be done to improve a person’s day to day quality of life.

Why have I been chosen?

Pam is seeking four people living with PCA and their carers to undertake a detailed case study analysis of the challenges they face day to day due to PCA.

Do I have to take part?

Taking part is totally voluntary. You must not feel that you should participate if you feel uncomfortable about doing so.

What will happen if I do take part?

Pam will visit you at your home (or other preferred location) and talk to you about your experiences. These conversations (with your permission) will be tape recorded. You will be free to stop these conversations at any time if you feel uncomfortable or distressed.

Will my taking part be kept confidential?

All information which is collected during the course of the research will be managed in accordance with the Data Protection Act. Audio recordings made during the assessment will be kept on a secure computer system at UCL and only listened to by research staff involved in analysing themes raised during your interview. All information about you will have your name and address removed so that you cannot be recognised from it. Information about you and your involvement in the study will be handled by the lead researchers (Dr Sebastian Crutch at University College London, Professor Nick Tyler of University College London and Professor May Gilhooly of Brunel University) and their team members. This information, including audio recordings and transcripts will be held for at least 10 years so that further ethically approved research may be conducted in the future.

What are the possible disadvantages and risks to taking part?

There are no risks involved in the interviews.

What are the possible benefits to taking part?

The benefits, we hope, will be in the future in helping to understand the brain and to develop aids and strategies which help one to better cope with problems regarding the environment and day to day living.

What happens if something goes wrong?

If you are unhappy with your treatment at any point during the research study, you are entitled to complain through the University College London Hospital Trust complaints procedure.

What will happen to the results of the research study?

The results will be presented to the scientific and medical community to improve further research. You are very welcome to request a copy of any resulting publications, and if you would like you may receive a copy of the Dementia Research Newsletter which describes this and other work taking part in this department.

Who is organising and funding the research?

This study is being organised and funded by the Alzheimer's Research UK, Economic and Social Research Council and National Institute for Health Research.

Who has reviewed the study?

This study has been reviewed by the London-Queen Square Research Ethics Committee.

Contact for further information

Please contact [REDACTED] or [REDACTED]
[REDACTED]

Thank you for considering taking part in this study.

You will be given a copy of the information sheet and a signed consent form to keep.

University College London Hospitals



NHS Foundation Trust

Dementia Research Centre
National Hospital for Neurology & Neurosurgery
Queens Square
London
WC1N 3BG
Telephone: 020 7829 8773

CONSENT FORM

Title of Project: *“A qualitative study exploring the specific everyday social and environmental challenges faced by community dwelling people living with posterior cortical atrophy (PCA)”*

Name of Researcher: Pam Turpin, University College London

Please mark each box with an X to indicate your agreement

1.	I confirm that I have read and understand the information sheet dated 28 th January 2016 for the above study and have had the opportunity to ask questions?	
2.	I understand that my participation is voluntary and that I am free to withdraw at any time, without giving any reason, without my medical care or legal rights being affected	
3.	I understand that an audio recording will be made and reviewed by research staff investigating how to live with dementia-related visual impairment. I understand my name and any other identifiable details will be removed from the recording.	
4.	I confirm I have had sufficient time to consider whether or not I want to be included in the study.	
5.	I agree to take part in the above study.	

Name of Participant

Date

Signature

Researcher

Date

Signature

Appendix Four: Topic guide

Topic Guide

<p>1. SETTING THE SCENE</p>	<ul style="list-style-type: none"> • Please tell me about yourself: • Age • Sex • Marital status • Any disabilities • How long married/together • Where you grew up and live • School/University/Career • Retired or not • Story of your diagnosis
<p>2. CHALLENGES</p>	<ul style="list-style-type: none"> • Tell me about when you realised you were having difficulties? <ul style="list-style-type: none"> ○ Explain the difficulties ○ What did you do ○ Who did you see ○ What help have you received • What medication do you take? • Do you have particularly difficulties coping independently? • What do you feel are the specific difficulties you face in communicating your needs to others? • What problems do you have with day to day activities? • Do you ever talk about your problems other than (name)? • Have you ever suffered from hallucinations?
<p>GENERAL CHALLENGES</p>	<ul style="list-style-type: none"> • What do you feel are the particular challenges you face day to day because of PCA? •

FOCUSING DOWN	<ul style="list-style-type: none"> • Now I would like to focus down on particular aspects of PCA
3.GENERAL HEALTH	<ul style="list-style-type: none"> • How is your general health? <ul style="list-style-type: none"> ○ Energy levels ○ Exercise ○ Mobility • Any other long-term conditions
4.MEMORY	<ul style="list-style-type: none"> • How would you describe your memory? <ul style="list-style-type: none"> ○ Good ○ Fair ○ Poor • Explain what you mean • How does this impact on you both
5.VISION	<ul style="list-style-type: none"> • Apart from PCA, do you have any other eye problems – short/long sight, cataract, macular degeneration, glaucoma etc • What are your particular vision problems • How much can you can see? <ul style="list-style-type: none"> ○ Do colours help or hinder? ○ Shadows ○ Contrasts ○ Patterns ○ Hallucinations ○ Perception • See the whole scene
6. BALANCE	<ul style="list-style-type: none"> • Do you have any difficulties with your balance? <ul style="list-style-type: none"> ○ Yes: in what way ○ No: ever? • What about walking – is that a problem? <ul style="list-style-type: none"> ○ Different surfaces? ○ Carpets – patterned or plain

	<ul style="list-style-type: none"> ○ Pavements
7. DISORIENTATION	<ul style="list-style-type: none"> ● Do you ever get lost? <ul style="list-style-type: none"> ○ In own home ○ In a room ○ New places ● How do you cope with this?
8. ACTIVITIES OF DAILY LIVING	<ul style="list-style-type: none"> ● Do you have any particular problems with: <ul style="list-style-type: none"> ○ Bathing – explain ○ Dressing - explain ○ Cooking – explain ○ Eating - explain ○ Socialising - explain ○ Shopping - explain ○ Driving – explain ○ TV, Radio, Reading ● THINK – RECOGNITION, HANDLING ● ASK – DO YOU HAVE ANY STRATEGIES THAT HELP? ● Have you had dealings with OCCUPATIONAL THERAPISTS ● Do any of the RNIB products help: <ul style="list-style-type: none"> ○ Liquid level indicator ○ Talking gadgets, watch, scales etc ○ Talking books
9. HEARING	<ul style="list-style-type: none"> ● Do you have any problems with your hearing? ● Yes – what kind? ● No – go on ● Have you ever had hearing hallucinations? ● How does this impact on you?
10. Physical Environment	<ul style="list-style-type: none"> ● Do any particular surfaces bother you? <ul style="list-style-type: none"> ○ Colour ○ Lines

	<ul style="list-style-type: none"> ○ Uneven surfaces ○ Colours ○ Equipment
11.PERSON	<ul style="list-style-type: none"> ● How does this situation impact on you? ● Anyone to help you? ● Stress levels/depression ● How has this situation impacted on your relationship with (name)? ● How do you view the future? <ul style="list-style-type: none"> ○ Positive ○ Negative
ENDING	<ul style="list-style-type: none"> ● Thank you for your time ● A copy of the transcript of our discussions will be sent to all participants to ensure you are happy with the contents. ● A copy of the findings for the research will be sent to you for your information.

Appendix Five: Example of NVIVO Nodes used in the analysis of the interviews

Node	Count	Date	Source	Date	
Touch	3	18/11/2017 15:02	PJT	18/11/2017	
Relationships	0	0			
With Spouse or partner	8	43	18/11/2017 15:15	PJT	15/01/20
With friends	2	5	18/11/2017 15:15	PJT	03/01/20
With family members	5	9	20/11/2017 10:53	PJT	08/01/20
Physical Environment	7	34	18/11/2017 15:04	PJT	15/01/2018
Indoors	4	13	30/11/2017 18:52	PJT	13/01/20
Outdoors	2	3	30/11/2017 18:52	PJT	04/01/20
PCA	8	26	19/11/2017 17:50	PJT	15/01/2018
Disorientation	1	2	18/11/2017 15:00	PJT	06/01/20
Activities of Daily Living	0	0	18/11/2017 15:01	PJT	18/11/20
Diagnosis	10	62	19/11/2017 17:51	PJT	15/01/20
First symptoms	11	61	19/11/2017 17:52	PJT	15/01/20
Identity	2	2	20/11/2017 10:49	PJT	12/12/20
Emotions	5	10	30/11/2017 18:50	PJT	19/01/20
Reaction when first diagnosed	5	8	03/12/2017 16:19	PJT	15/01/20
Falls	9	37	03/12/2017 16:35	PJT	15/01/20
Visual problems	12	64	11/12/2017 10:36	PJT	15/01/20
Carers day to day life	4	27	11/12/2017 14:23	PJT	13/01/20
Support	4	12	11/12/2017 14:24	PJT	15/01/20
protecting	4	9	11/12/2017 14:35	PJT	15/01/20
Hiding the truth	1	2	11/12/2017 14:40	PJT	11/12/20
Opinions of others	7	19	11/12/2017 14:41	PJT	15/01/20
Coping strategies	2	3	11/12/2017 14:43	PJT	06/01/20

Appendix Six: Presenting Symptoms of each interviewee living with PCA

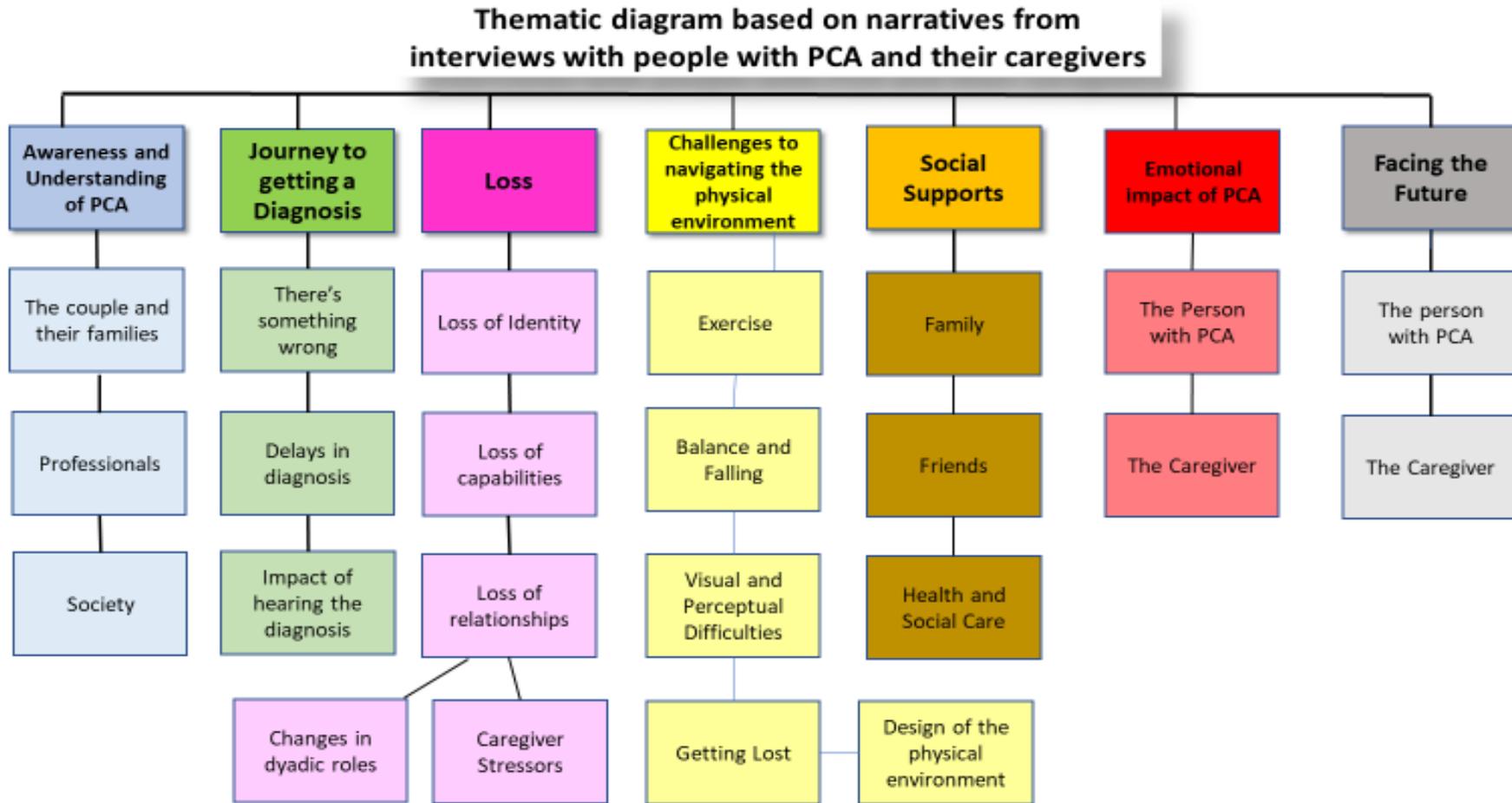
Symptoms of PCA*	Betty	Polly	Brian	Laura
Stage 2				
a loss of confidence in their driving ability	✓	✓	✓	✓
problems using a computer	✓	✓	✓	✓
difficulties with reading, writing, spelling and/or arithmetic	✓	✓	✓	✓
difficulty recalling and writing down telephone numbers	✓	✓	✓	✓
difficulty assembling novel devices/objects	✓	✓	✓	✓
The person may experience vertigo or other balance problems	✓	✓	✓	✓
may experience difficulties finding the right words when speaking or may experience odd visual sensations (e.g. occasional changes or washes of colour in the centre or periphery of their vision)	✓	✓	✓	✓
Stage 3				
Difficulty seeing things or only seeing part of something	✓	✓	✓	✓
General 'clumsiness'	✓	✓	✓	✓
Difficulty finding things in a handbag, cupboard, etc. –	✓	✓	✓	✓
Difficulties dressing and performing other activities of daily living that require spatial judgement	✓	✓	✓	✓
Inability to tell the time from a clock or watch	✓	✓	✓	✓
Reading and writing difficulties	✓	✓	✓	✓
Mild memory problems for recent events	✓	✓	✓	✓
Mild difficulty retrieving the correct word in conversation	✓	✓	✓	✓
Stage 4				
Inability to read labels on food packets –	✓	✓	✓	✓
Problems discriminating coins –	✓	✓	✓	✓
Inability to tell the time from an analogue or even digital watch	✓	✓	✓	✓
Dissociation in ability to walk, sit down and stand up				✓
Being able to see some things but not others	✓	✓	✓	✓
Cannot read but can still follow or at least get pleasure from TV or the cinema	✓	✓	✓	✓
Getting lost within the home.	✓	✓	✓	✓
An inability to identify even very familiar faces				✓
Resorting to eating with one's hands, or being fed	✓	✓		✓
increasing word finding difficulties	✓		✓	✓
Stage 5				
Require help with most or all everyday tasks, including toileting and personal care	✓	✓	✓	✓
Experience spatial problems not limited to vision	✓	✓	✓	✓
Require support and/or supervision when walking	✓	✓	✓	✓
Episodic memory and word finding difficulties may also now be prominent			✓	✓
Some people may have problems with multiple senses				
Increased sensitivity to pain		✓		
become more sensitive to the cold	✓	✓	✓	✓
Sense of hearing may change				
Feelings of imbalance or instability	✓	✓	✓	✓
Stage 6				
Experience changes in sleep patterns			✓	✓
Have increasingly frequent trouble controlling their bladder or bowels –				✓
Become quiet and withdrawn				✓
Experience behaviour and personality changes		✓		✓
Have difficulty communicating				✓
Experience continued deterioration of sensory functions				✓
Stage 7				
Loss of ability to respond to their environment				
Unable to carry on a conversation				✓
Unable to control movement.				
Total functional dependence				
Unable to walk				

*as described by (Crutch, Walton, Carton, & Shakespeare, 2017)

Appendix Seven: International Consensus Research Diagnostic Criteria

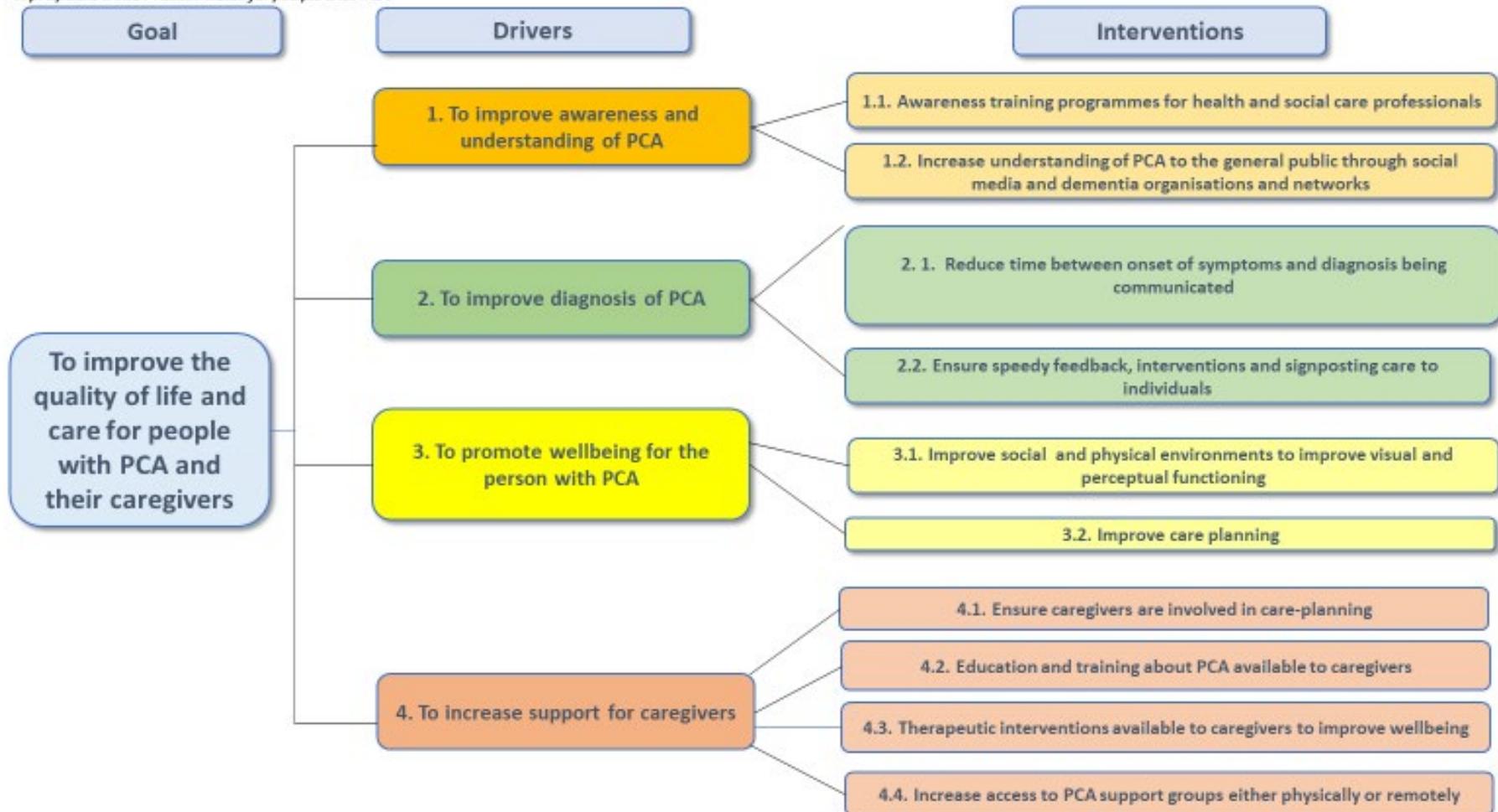
<p>Appendix Seven : International consensus research diagnostic criteria for posterior cortical atrophy</p>
<p>Clinical Features:</p> <ul style="list-style-type: none"> • Insidious onset • Gradual progression • Prominent early disturbance of visual function and/or other posterior cognitive features
<p>Cognitive features:</p> <p>At least three of the following must be present as early or presenting features and were the predominant features initially impacting activities of daily living:</p> <ul style="list-style-type: none"> • Space perception deficit • Simultanagnosia • Object perception • Constructional dyspraxia • Environmental agnosia • Oculomotor apraxia • Dressing apraxia • Optic ataxia • Alexia • Left/right disorientation • Acalculia • Limb apraxia • Prosopagnosia • Agraphia • Homonymous visual field deficit • Finger Agnosia
<p>All of the following must be evident:</p> <ul style="list-style-type: none"> • Relatively spared anterograde memory function • Relatively spared speech and nonvisual language functions • Relatively spared executive functions • Relatively spared behaviour and personality
<p>Neuroimaging:</p> <ul style="list-style-type: none"> • Predominant occipito-parietal or occipito-temporal atrophy (MRI) or hypometabolism (FDG-PET) or hypofusion (SPECT)
<p>Exclusion criteria:</p> <ul style="list-style-type: none"> • Evidence of a brain tumour or other mass lesion, cerebrovascular disease, afferent visual cause or other identifiable causes for cognitive impairment (e.g. renal failure)
<p>(Crutch S. J., et al., 2017) page 876</p>

Appendix Eight: Thematic diagram based on narratives from interviews with people with PCA and their caregivers



Appendix Nine: Diagram of proposed Good Practice Guide for people with PCA

A proposed Good Practice Guide for people with PCA



Appendix Ten: Flow Diagram showing links between the Themes and the Good Practice Guide Drivers

