

‘It’s building up a picture that’s the problem’

Posterior cortical atrophy is one of the rarer types of young onset dementia, initially affecting vision while often leaving memory intact. **Teresa Nahajski** and **Helen Scott** set out a collage of carers’ comments giving an insight into the condition



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Young onset dementia (YOD) accounts for about 5% of the total number of people estimated to have dementia in the UK, the most common types of YOD being Alzheimer’s and vascular dementia. But there are also some rarer YOD variants which have less well known symptoms, including posterior cortical atrophy (PCA), which differs from other dementias in leading to progressive visual and spatial dysfunction.

As its name suggests, PCA affects the posterior cortex of the brain, the parietal and occipital lobes. Initial symptoms are often associated with visual, literacy and numeracy skills while memory and insight may remain unaffected until the later stages (Carton et al 2015). Difficulties with reading, writing and dealing with numbers are prominent aspects.

But even while losing these abilities, people with PCA can remain verbally articulate as the late author Sir Terry Pratchett demonstrated. He raised public awareness of the condition when he announced his diagnosis in 2007, at the age of 59, and described how he continued to write books even though the symptoms affected his ability to spell and read words in print (Alzheimer’s Research UK 2013).

Other visual and spatial symptoms of PCA can be difficulties with balance and judging distances, recognising once familiar items and experiencing odd visual sensations such as changes in colour or blind spots. In the later stages, PCA can lead to more severe difficulties with vision and balance which impact on mobility (Carton et al 2015).

There has been little research into the psychological impact of PCA on those diagnosed and those who care for them. The qualitative study outlined here aimed to explore the experiences of living with a partner who had been diagnosed with PCA, looking in particular at the transition from partner to carer and the coping strategies that were employed by both parties as the condition progressed.

Because of the rarity of PCA and the qualitative nature of the research, just six carers were recruited from across the UK. Semi-structured interviews were carried out which were transcribed and analysed to identify themes. For three of the participants, their partners had reached the stage of needing full time care, either in care homes or at home, whereas in the

remaining three cases their partners were still in the earlier stages of the condition.

All participants talked about their experiences with honesty and frankness, saying that they wanted to share their stories and accumulated knowledge with professionals, carers and others who found themselves supporting someone with PCA.

Initial impact

Participants were asked to describe what the first indications were that something was unusual or wrong, and how this led to their or their partner’s decision to seek medical advice, usually via ophthalmologists or their GPs. Here are two typical responses:

Things were happening so slowly we were adapting without really realising. There were some signals I can see with hindsight. F got lost twice in places he would have known very well, but they were over quite a long period – that’s why I didn’t connect them.

A few years ago, B told the optician “I know it sounds silly but sometimes my eyes feel funny”. So she referred him initially to the eye hospital.

Changes in visual perception and spatial awareness can be very gradual as they begin to have an impact on everyday life. Difficulties arose with some tasks but not others:

We noticed when A was pegging out washing, he might have four pegs on one item of clothing and then they might go across three lines. However, he’s still very articulate, and he is highly conscious of all the things he can no longer do.

Physical coordination could be affected as the person with PCA might not accurately interpret what they saw around them and found that objects became difficult to distinguish. The unpredictable nature of the symptoms could compromise activities and hobbies outside the home, as these two participants illustrate:

When B was playing golf, he could hit the ball, but then he would be looking around for it and he couldn’t see it. And then he’d look down and it would come into view, but by the time he’d chosen his club he couldn’t see it again!

A number of dementia support agencies exist, but carers do not always have the time or knowledge to research what is available locally. Study participants suggested that it would make sense to have one point of contact for a centralised support network

When A's walking he will walk diagonally across the road, he won't see the shortest way across. And at crossings, he'll get the green traffic lights muddled with the green man, so he'll start to walk across when it's green traffic lights!

Following instructions

Understanding and processing instructions that involve recognition of objects or visualisation often became problematic. Reading and writing could cause difficulties in this respect, for example, not being able to distinguish patterns in larger print or follow lines of text across a page:

C would often ask me where something was simply because it was out of her line of sight. It's like the children's game "peek-a-boo": the person disappears behind the chair and they've gone. It's like her world is just a two-dimensional thing in front of her and she has to think really hard to think something might exist if it isn't in front of her.

It's building up a picture that's the problem. So if a letter is too near and too big, they can't work out what it is, because the eye has to scan around it and build it up into an image.

Impact on independence

The ability to drive safely was often one of the first skills to deteriorate, given that problems with spatial awareness could result in poor judgement in relation to distance. If the partner had to give up driving, this could be the first perceived crisis point. Both partner and carer could lose a degree of independence and access to social activities, leading to isolation.

At first, F was safe to drive, he didn't go very far anyway and I suspect he was restricting himself. But when they said he wasn't fit to drive, that's really when he couldn't deny it any longer.

Problems understanding directions could make

it hard to get to the toilet in time when out and about, which again had a detrimental effect on activities outside of the home. For younger people with no physical signs of disability, this could make social situations difficult. Partners were still able to walk, of course, so they needed alternative evidence of incapacity to obtain access to some disabled facilities such as disabled parking.

C can walk fine, it's just that she can't walk anywhere in particular. You can't describe to her where the toilet is if it's round two corners, because she wouldn't be able to keep track of what you're talking about.

It became quite difficult to go out because I was worried if I let D go to the ladies, would she find the way in, would she sort herself out? I'm not sure she was incontinent, it was more about her not being able to find and use the toilet.

For those still in work, any deterioration in skills was often covered up or denied by the person with PCA until it was severe enough to affect their ability to cope with their workload. This could eventually lead to loss of their job:

They put E on lots of different medication and from that point he just lost it completely. He couldn't drive, he didn't know where he was, he didn't know which direction to go in. It was as if he was looking at stuff but couldn't see it.

Practical support

In terms of practical support, research has been carried out into simple adaptations that could help to compensate for the visual impairment, such as simplifying the environment, ensuring good lighting, and using talking clocks and other assistive technologies (Carton *et al* 2015). These could prolong the person's ability to read, helping to maintain their quality of life (Crutch *et al* 2016). Further research and development of these technologies would be useful for those adapting to the visual problems of PCA.

Support groups for people with rare dementias have started up and these can be valuable in sharing experiences, teaching new skills and preparing people to cope better and for longer. Fear, anxiety and isolation are correspondingly reduced (Carton *et al* 2015). A number of dementia support agencies exist, but carers do not always have the time or knowledge to research what is available locally. Study participants suggested that it would make sense to have some form of centralised support network with one point of contact.

Raising awareness of PCA and other rare forms of dementia can only help in solving the practical and social problems people face, especially in the later stages of the condition. For carers, contact with others in similar situations would allow them to share problems and ideas to resolve them.

In future, more support groups, dementia cafes and meeting centres for the rarer dementias will have an important role to play in managing the practical and emotional impact of these conditions. ■

References

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