Posterior cortical atrophy: understanding and support

Posterior cortical atrophy (PCA) is an unusual form of dementia that primarily affects sight rather than memory. In the first of two articles, Amelia Carton and colleagues describe PCA and suggest practical ways to respond.

Posterior cortical atrophy (PCA) is a progressive neurodegenerative syndrome characterised by a decline in visual processing associated with loss of cells in the parietal and occipital lobes at the back of the brain (Crutch et al 2012). In the PCA variant of dementia, the visual parts of the brain fail to correctly interpret the information gathered by the healthy eyes about the surrounding world. One way to put it is that PCA is a problem of ‘brainsight’ not eyesight.

Although impairments in visual processing are most characteristic, other skills dependent on posterior brain regions are also often affected including literacy, numeracy and praxis. PCA is most commonly caused by Alzheimer’s disease pathology (AD) (Renner et al 2004). However, in contrast to individuals with the typical memory-led form of AD, episodic memory and insight are relatively well preserved in PCA.

Instead, often the first symptoms noticed include difficulties with driving and fine spatial judgements, misperceiving what or where objects are, and deterioration of reading, writing, spelling and counting abilities. The reasons why Alzheimer’s disease pathology can affect different people’s brains in such different ways (the memory centres in one person, the vision centres in another) are not yet fully understood.

Path to diagnosis

PCA is also typically a young-onset condition with the first features usually presenting in the 50s or early 60s (McMonagle et al 2006; Mendez et al 2001). As with other young-onset dementias, this means that when symptoms are first experienced many people are in paid employment, have young or teenage children living at home, are physically fit and healthy, and have significant financial and other commitments, such as mortgage responsibilities or caring for an older relative or parent. The combination of an early age at onset and an unusual physical manifestation or phenotype may also contribute to the particularly slow path to diagnosis experienced by many people with PCA, whose early symptoms may be mistakenly attributed to impaired eyesight, anxiety, depression, menopause or malingered.

Main features of PCA

As with any diagnosis, people with PCA and their families often want to manage their expectations about their diagnosis by forming a knowledge base of how the condition is likely to progress. This extra information might enable them to better plan for the future, to make any necessary arrangements such as for support or finances, and also to anticipate and adjust to new symptoms, changes or challenges. A recent guidance document entitled The Stages of PCA (see ‘Resource’ below) provides a general framework describing the common stages of the condition in an effort to better equip those affected with an understanding of how the condition might progress. It reflects the different symptoms, deficits and challenges faced by individuals diagnosed with PCA relative to more typical, memory-led presentations of Alzheimer’s disease.

Common features and problems described in the mild stages of PCA (Stages 2-3) include:

- a loss of confidence when driving, using escalators and so on, because it is harder to judge distances
- experiencing vertigo or other balance problems (although some aspects of these may develop later or not at all)
- experiencing odd visual sensations (eg occasional changes or washes of colour in the centre or periphery of their vision)
- difficulty seeing things which are ‘right under your nose’ or only seeing part of something (eg not noticing food on the edge of a plate)
- difficulties dressing and performing other activities (eg DIY, cutting a piece of toast in half) that require spatial judgement (eg using stairs) and coordination (eg left-right choices)
- getting lost on the page when reading
- people at this stage may also experience subtle sensory changes and disturbances (eg feeling cold)
- anxiety, often as a result of insight into these experiences coupled with uncertainty as to the cause. Anxiety and feeling flustered can exacerbate the visual and other problems.

These stages may reflect the earliest signs of PCA, especially where eye tests reveal no indication of ophthalmological problems. Problems more commonly observed in the moderate stages of PCA (Stages 4-5) include:

- dissociation in ability to walk, sit down and stand up (eg 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 (eg following moving targets better than localising static objects; can’t read but can still follow or at least get pleasure from TV or the cinema)
- despite knowing that they are at home, people with PCA may have difficulty navigating between rooms or locating particular places (eg the bathroom)
- an inability to identify even very familiar faces, with recognition of identity dependent on hearing someone’s voice
- difficulty manipulating cutlery and seeing the location of food on a plate
- viewing the world 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
• difficulty positioning the body relative to furniture and seeing and feeling the position of clothing
• experiencing spatial problems not limited to vision, with spatial commands and actions (eg ‘turn round’, ‘shuffle forwards’) difficult to comprehend and execute
• many individuals are registered as blind or partially sighted, meaning they require support in all visually-guided activities
• difficulty sensing the relative position of parts of the body (as pronounced as not knowing if one is ‘the right way up’)
• increased sensitivity to certain sounds.

In the more severe stages (Stages 6-7) the experience of PCA may mirror that of typical AD more closely than in the earlier stages of the condition, although vision remains the most pronounced impairment with most people being diagnosed as functionally blind.

Practical aids and advice
People with PCA have been shown to experience significantly greater impairments in everyday skills and self-care than people with typical AD, and by contrast less impairment in aspects of memory, orientation and motivation (Shakespeare et al 2015). Consequently, much of the advice recommended for and by people with PCA focuses on practical strategies and aids (see box).

Support groups
Support groups have a constructive role to play in offering people the opportunity to acknowledge their diagnosis and its consequences alongside peers in a similar position who understand their predicament (Suarez-Gonzalez et al 2015). Naturally, the benefits of such support are not unique to PCA. However, there is lower recognition and understanding of the symptoms and consequences of the condition, and owing to the relative rarity of the diagnosis many people are left asking, “Is it just me?” Additionally, evidence suggests that caregivers of younger people with dementia have higher levels of burden than their older counterparts, even when matched for severity of dementia and behavioural disturbance (Freyne et al 1999).

These factors provided the motivation for the formation of the first PCA Support Group in 2007 (www.ucl.ac.uk/drc/pcasupport). The group offers face-to-face meetings which provide an opportunity to meet with and learn from clinicians,

Common strategies
Asking people to introduce themselves before starting a conversation
Using plates which contrast with the colour of the food being eaten, or eating food which does not require cutlery
Buying clothes with poppers, rather than buttons/shoes without shoelaces
Simplifying the environment by removing clutter to keep pathways clear, ensuring lighting is adequate and by reducing glare (e.g. installing curtains to block direct sunlight and avoiding using light bulbs without shades)*
Placing stickers on to glass doors and windows to make them more noticeable*
Using bumpers or dots to mark important areas (eg dishwasher settings, microwave or stove dials)*.

Common aids
Audible or vibratory liquid level indicators to indicate when a cup is full
Talking clocks/watches
Mobile telephones with pre-programmed dialling and simplified displays, tactile markers to help identification of buttons
Audiobooks
Chopping board with white side and black side to increase contrast while cutting*
PenFriend audio labeller
A RADAR key to access public toilets for disabled people.


researchers and most importantly other people with PCA and their carers, families and friends. The social and emotional benefits of participating in such a group are facilitated outside formal meetings through telephone contact networks, social media, contributions from group members in regular newsletters, and increasingly through carer-led local meetings. Sharing life-changing experiences in a safe environment, together with learning new skills and coping strategies and preparing for the future, can enable people to cope better and for longer and lead to a reduction in fear, anxiety, and isolation.

Individual support
There has been little empirical work on psychotherapeutic approaches to mental health problems in PCA or young onset dementia more generally. Experience from the psychological therapies service attached to the specialist cognitive disorders clinic at the National Hospital for Neurology and Neurosurgery in London suggests that CBT techniques can be used with very little modification with patients with PCA, because they demonstrate relatively preserved insight and are usually capable of holding, understanding, and manipulating information in the mind (though may understandably need assistance with written materials). Psychoeducation about PCA is also key for patients, carers and families.

Common themes arising in therapeutic sessions with people with PCA that seem less prominent for other forms of dementia include comparatively very early loss of skills necessary for privacy and independence, such as dressing and reading, accompanied by acute awareness of that loss. People may feel guilty, angry, frustrated and low at becoming so dependent on others for basic functions. Misperceptions are another distinguishing theme, allied to a lack of understanding of non-memory symptoms and discrepancies between lost and preserved skills. For example, someone may not be able to reach out and pick up a mug of tea because of characteristic difficulties in seeing where or what things are, whereas they are still able to converse articulately about current affairs and remember recent events. Such misperceptions can lead to conflict and psychological distress in both patients and caregivers.

Conclusions
Naturally, no description of a condition can entirely capture the experiences and difficulties of all individuals with that diagnosis. However, by including a broad description together with experiences from patients and carers and brief summaries of strategies, aids and models of individual and group support, we hope this article captures something of the essence of living with PCA.

To appreciate the wide-ranging implications of the diagnosis, it is important to recognise both its core, near-universal features (eg visual problems), but also a broader range of symptoms which might occur as it progresses (e.g. balance and other sensory changes). It is also hoped that learning about the pronounced visual problems experienced by people with PCA will also raise awareness about the more subtle and under-recognised dementia-related visual problems which eventually affect the vast majority of people with more typical Alzheimer’s disease and several other dementias. •
In the second article of this series of two, we will explore further the quantity and quality of the information on PCA readily available to health and social care practitioners.

References

Acknowledgements
This work was undertaken at UCLH/UCL, which received a proportion of funding from the National Institute for Health Research (NIHR) biomedical research centres funding scheme. The Dementia Research Centre is an Alzheimer’s Research UK (ARUK) co-ordinating centre. The work was supported by the NIHR Queen’s Square Dementia Biomedical Research Unit, and by an ARUK senior research fellowship and Economic and Social Research Council / NIHR grant. Jill Walton and the PCA Support Group are supported by the Myrtle Ellis Fund and anonymous donors.

Resource
The Stages of Posterior Cortical Atrophy (PCA) is written by Sebastian Crutch, Jill Walton, Amelia Carton and Tim Shakespeare at Dementia Research Centre, UCL Institute of Neurology, in association with members of the PCA Support Group. It is available by clicking on “Stages of PCA” at www.ucl.ac.uk/drc/pcasupport and is written in a language common to healthcare professionals and families. The document is based on the well-established ‘7 Stages of Alzheimer’s Disease’ framework, derived from Reisberg and Franssen’s Clinical stages of Alzheimer’s disease (Reisberg & Franssen 1999).

Care and compass
A plethora of new safeguarding policies has failed to prevent neglect and abuse, say Lynne Phair and Hazel Heath. In the first of three articles, they call for a new emphasis on the “golden thread” of compassion.

Adult safeguarding is gradually being enshrined in law in every part of the UK.* There is now a plethora of adult safeguarding policies, guidance and research amid growing concern about standards in health and social care, fuelled by the findings of the public inquiry into the Mid Staffordshire NHS Foundation Trust (Francis 2014) and the serious case review of Orchid View care home (Georgio 2014).

Despite all these new laws and policies, as Eastman (2015, p4) observes, “still far too many people experience neglect, treatment bordering on abuse and a total lack of compassion”. One of the reasons behind this, he argues, is that “too frequently there is a disconnect between policy intention and practice. It is one thing to be told that safeguarding duties apply to an adult who needs care and support, who is experiencing or is at risk of abuse or neglect, and who as a result of those needs is unable to protect themselves; it is quite another to provide coherent organisational systems and individual practice responses.”

Eastman concludes that one ‘golden thread’ needs to be reinforced and strengthened more than ever at every level of our care commissioning, governance, management, provision and practice: compassion. This article, the first in a series of three, explores how we can focus on compassion in adult safeguarding through individual care practice in care homes. The second article will focus on adult safeguarding through the promotion of compassion in care cultures and the third looks at leadership and education/training.

Some principles in adult safeguarding are acknowledged as context.

Principles in safeguarding
Analysis of the principles underlying adult safeguarding legislation and guidance suggests that, despite some local differences, these are broadly similar across the UK. For example the Care Act 2014 in England and the accompanying statutory guidance establish six principles of safeguarding:

• Empowerment: There is a presumption of person-led decisions and informed consent.
• Protection: There is support and representation for those in greatest need.
• Prevention: It is better to take preventative action before harm occurs.
• Proportionality: There is a proportionate and least intrusive response appropriate to the risk presented.
• Partnership: Local solutions are created through services working with their communities. Communities have a part to play in preventing, detecting and reporting neglect and abuse.
• Accountability: Accountability and transparency in delivering safeguarding.

While all of these principles are important, prevention is paramount in that good anticipatory procedures and practices can reduce the risk of neglect and abuse, as well as stopping the development of abusing cultures of care. Activity for safeguarding, along with training, education and leadership, should use a model that will support prevention, empowerment and protection, while ensuring the resident remains central to all activities. Phair’s model (see Phair 2015 for details) focuses on prevention, learning from others, investigation, and practice and policy development.

Principles, guidance and models of good practice are, of course, important but, as Eastman emphasises, these must not drive our work and interventions. “Instead, kindness and compassion should underpin our professional and organisational responses within the