Expert essay

Posterior cortical atrophy

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Posterior cortical atrophy (PCA) is a neurodegenerative syndrome characterized by predominant and progressive loss of higher order visual and other posterior cortical functions consistent with parieto-occipital and occipito-temporal involvement, despite relatively preserved memory, language, and insight. PCA tends to have a young onset presentation, typically around 50–65 years, being underpinned by Alzheimer's disease (AD) pathology. Core features of PCA include space and object misperception, features of Balint's and Gerstmann's syndrome, apraxia, environmental agnosia, diminished reading and face perception and visual field defects (Table 1[1]).

Table 1

Clinical features
Space and/or object perception difficulties
Simultanagnosia, optic ataxia, and oculomotor apraxia
Dyscalculia, dysgraphia, left-right confusion, finger agnosia
Constructional, dressing, and/or limb apraxia
Environmental agnosia
Reading difficulties
Face perception difficulties
Relatively spared anterograde memory, speech, non-visual language, executive function and behaviour

Early PCA symptoms include difficulties with driving, reading, dressing, and orientating to familiar environments. PCA symptoms often carry significant implications for autonomy and personhood; feelings of disempowerment and depression may arise in individuals unable to use a telephone or remote control despite spared insight[2].

Common misconceptions and clarifying needs

Consistent with individuals with other young onset and atypical forms of dementia, people with PCA have often experienced a lengthy period of uncertainty before diagnosis (Table 2[3]). Stress associated with this often-convoluted diagnostic journey may be exacerbated by challenges regarding employment, finances and changing family roles and the need to frequently re-explain PCA owing to limited public and professional awareness[2].

Table 2

Diagnostic red flags
Repeated appointments with eye specialists
Repeatedly changing prescription of glasses
Misdiagnosed with ocular condition
May undergo unnecessary surgeries (e.g., cataract removal)
May be diagnosed as having a functional disorder

Factors complicating PCA diagnosis may cause continuing frustrations and undermine subsequent support provision. For example, the easy misattribution of PCA cortical visual problems as eyesight rather than 'brainsight' issues may prompt support designed to assist with sensory but not concurrent cognitive decline. A related misperception limiting the effectiveness of post-diagnostic support is the notion that PCA is solely a 'visual dementia'. Perceptual ('what?') and spatial ('where?', 'how?') changes may manifest not only through disturbed visual, but also auditory and somatosensory functions[3][4][5] visual imagery and spatial concepts like left and right[1]. Awareness of non-visual deficits is critical to design, tailor and communicate support strategies with, for and to the individual with PCA and their care partner.

Those caring for a person living with PCA may be surprised by variations in visual functions, from inconsistent visual field testing to dissociations between single word and space perception[3][6]. Variable field deficits can result in apparently implausible difficulties with misperceiving objects presented in clear view (albeit in inferior peripheral vision) despite better perception of objects presented at a distance, or misreading newspaper headlines despite better reading of small print. These inconsistencies challenge not only understanding of symptoms, but also confidence in carrying out superficially simple tasks which have become more effortful and unreliable.

Post-diagnostic support, information, and management

Prominent visual, motor, language and other symptoms carry particular implications for self-care, social and leisure activities for persons living with PCA and their carers and support needs (Table 3). Most persons living with PCA will not be safe to drive, with consequent ramifications regarding dependency. Providing tailored information, advice and support regarding PCA symptoms and everyday difficulties, time-sensitive aids and adaptations to manage symptoms, and psychosocial interventions may improve functional status and wellbeing.

Table 3

Considerations
A key priority is discussion of driving safety; most individuals will not be safe to drive
Occupational and daily routines may be severely impacted by progressive cortical visual loss, despite relatively preserved memory, language and insight
Individuals may have a high risk of becoming lost
Individuals may be eligible to register as severely sight impaired or blind, even despite normal visual acuity
As PCA progresses, most individuals will become functionally blind leading to a high risk of falls

Registration as partially-sighted or blind may facilitate access to relevant care and support services and financial and legal benefits[3]. Access to information and advice reflecting lived experiences of PCA is critical given limited public and professional awareness[2]. Co-developed resources include the Stages of PCA, describing symptom progression, public engagement, and professional development materials to improve recognition and management of PCA symptoms.

Pharmacological approaches

In principle, most individuals with PCA caused by AD should benefit from symptomatic or eventual disease-modifying treatments for 'typical' AD. In practice, few existing pharmacological therapies have been directly examined in PCA, and the inclusion or exclusion of PCA participants into conventional trials remains somewhat inconsistent.

Clinical benefit of cholinesterase inhibitors for PCA has been reported. While there have been no pharmacological trials in relation to management of PCA motor symptoms, expert review of corticobasal syndrome found limited support for pharmacological management of tremor, levodopa for rigidity and bradykinesia, and botulinum toxin injections managing dystonic spasms and pain[7]. Standard pharmacologic treatments may be considered for patients with persistent low mood or anxiety and recommending treatments for parkinsonism, seizures or myoclonus may be appropriate for individual patients[3].

Non-pharmacological approaches

Aids and adaptations

As with all dementias, advice regarding aids and strategies must be tailored to the individual, their condition (type and severity) and environment (social and physical), ideally with the involvement of a multidisciplinary clinical and/or support team. Practical tips for patients with PCA and their families have been collated based on occupational therapy and neurological practice (Table 4). Several compensatory approaches have been developed to mitigate PCA symptoms, particularly space/object perception, alexia, and environmental agnosia. Evidence of reducing visual clutter, increasing spacing and maximizing contrast supporting visual functions in PCA participants has informed the development of computer-based reading aids[6,8] and environmental adaptations comprising visual cues and lighting interventions to support navigation and walking[9,10]. Occupational therapists, speech and language therapists and physiotherapists may promote functional status and communication through compensatory approaches, communication skills training and managing falls[3][7].

Advice, social context, and peer-to-peer support

Beyond the individual, support and advice must be tailored to setting and context. Persons living with PCA face numerous barriers to service use and participation, with general dementia group activities often inappropriate due to age and/or symptom profiles, and psychosocial therapies relying on visual formats (e.g., reminiscence photographs) or advising against inclusion of people with visual impairment (e.g., cognitive stimulation therapy). During the COVID-19 pandemic, mask wearing, digital/video-based communication and social distancing posed particular challenges to those with praxic, proprioceptive, face, object and space perception and mobility difficulties.

Home-based interviews and observations with persons living with PCA and their care partners highlight the challenges and importance of maintaining ongoing engagement in daily activities which are both 'fun' (e.g., adapted artistic and music-related activities) as well as 'functional' (e.g., household chores and personal care). Findings also suggest the enduring importance of enabling persons living with PCA to continue to contribute to everyday life and to provide

Table 4

Non-pharmacological treatment
Individuals may benefit from referral to an occupational therapist, ideally with experience in supporting individuals with cortical visual loss, to develop compensatory strategies to support functional status and promote participation in meaningful activities (e.g. utilising voice-activated music listening devices)
There is evidence that reducing clutter may promote reading function[6,8]. Professional recommendations include simplifying the environment (e.g. removing clutter and unused objects), though with sensitivity to the potential emotional impact of removing objects which relate to an individuals' identity and personhood (e.g. books for a previously avid reader, tools for a former handyman)
There is evidence that strategic use of contrast, minimizing lighting variability and shadows may facilitate visually-guided navigation and walking[9,10]. Shared strategies from individuals include brightly coloured stickers to make parts of garments or buttons on gadgets more visually salient and motion-sensor lights or nightlights to support wayfinding to the bathroom
Use of a white cane or Sunflower lanyard may be helpful, particularly to encourage awareness of the individual's support needs among others in public places
While equipment designed for those with low vision might be appropriate beyond the white cane (for example, talking watch, typoscope, audiobooks), careful appreciation of concurrent non-visual symptoms is required. Diminished praxis skills and non-visual spatial awareness subsequently accompanied by declining memory and executive functioning pose substantial challenges to the adoption of generic assistive technology[3–5]

care and support for others, as well as receiving support for themselves – e.g., via long-established family or work roles – and a particular impact on identity given relatively intact long-term memory[3].

Given the symptoms, typically young onset presentation and context, syndrome-specific peer-to-peer support may be especially valuable for persons living with PCA. Syndrome-specific groups may provide opportunities for advocacy and sharing of strategies. Tailored peer support is available through formal multicomponent (mixed peer and professional) support services (e.g., Rare Dementia Support, Colorado PCA Support) and informal peer-led social media groups.

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