

Posterior cortical atrophy

This help sheet has been developed to assist your understanding of posterior cortical atrophy and its symptoms, diagnosis, progression and treatment.

What is posterior cortical atrophy?

Posterior cortical atrophy (PCA) is a progressive degenerative condition involving the loss and dysfunction of brain cells particularly at the back (posterior) of the brain. Brain tissue in the outer layer (cortical) shrinks (atrophy) as cells are lost. In the vast majority of cases, this loss of brain cells is associated with the same pathological brain changes seen in typical Alzheimer's disease, namely amyloid plaques and neurofibrillary tangles. In other words, PCA is most usually considered to be an unusual or atypical variant of Alzheimer's disease.

Differences to typical Alzheimer's disease

Despite being caused by the same disease process, the effects of PCA and typical Alzheimer's disease on the behaviour, thinking and skills of individuals with each condition can be very different. Typical Alzheimer's disease is most commonly associated with deterioration in memory, language, perception and other skills and abilities. By contrast, individuals with PCA tend to have well preserved memory and language but instead show a progressive, dramatic and relatively selective decline in visual processing and/or literacy skills such as spelling, writing and arithmetic.

The reason for this marked distinction between PCA and typical Alzheimer's disease is that the distribution of the disease in the brain is different. In typical Alzheimer's disease, disease-related changes are evident across most brain areas, whereas in PCA the changes are restricted to or focussed on the back of the brain and so initially only the skills which those brain areas normally support are affected.

Symptoms of posterior cortical atrophy

Difficulties with skills such as literacy, numeracy, and the ability to make skilled movements may be among the first symptoms noticed by individuals with PCA. Such difficulties may be experienced in the following ways:

- Problems recalling the exact spelling of words
- Difficulties with handwriting or typing
- Difficulties with remembering the shape or name of particular letters or numbers
- Slowness and difficulty with mental arithmetic
- Problems dealing with money and small change
- Awkwardness making gestures (for example, waving, thumbs up)
- Difficulties with using particular tools, kitchenware or implements (for example, cutlery, scissors, glasses)
- Problems with dressing and clothing (partly related to difficulties with visual perception)

Another common problem noticed by individuals with PCA is with visual processing. Quite understandably, this leads many people to consult an eye specialist, but in fact the visual problems experienced are not related to the eye. Rather, the problems stem from the difficulty the affected brain has with interpreting the information sent to it by the healthy eyes. The precise nature of the visual problems experienced may vary widely but often include difficulty with some or all of the following:

- Recognising objects in pictures (particularly if taken from obscure angles or incomplete)
- Recognising faces (for example, television characters, friends, relatives)
- Appreciating the spatial location of objects (for example, missing when reaching out to pick something up)
- Judging speed or distance (for example, when driving a car or walking down stairs)
- Perceiving movement among things which are stationary
- Following text when reading (for example, missing some lines of text)

- Reading particular words (for example, finding that letters appear to move around)
- Reading certain types of text (for example, large print such as newspaper headlines)
- Experiencing objects as having an unusual colour
- Experiencing increased sensitivity to bright light or shiny surfaces
- Seeing clearly (for example, experiencing double vision, or feeling that one's eyes are jerking around)

PCA can affect people in different ways initially. In some instances, the disease affects both sides of the brain equally, leading to a combination of many of the symptoms described above. For other people, the disease affects one particular brain area earlier or more significantly. As a result, problems with spelling and writing, for example, might be the first sign of the condition with vision relatively unaffected, whereas for others, difficulties in seeing where objects are might be the initial symptom.

Diagnosis of posterior cortical atrophy

PCA tends to affect people at an earlier age than typical Alzheimer's disease, with individuals often being in their mid-fifties or early sixties at the time of their first symptoms. However, it can also affect older people. The first signs are often subtle symptoms which may be difficult for the person experiencing them to explain. Individuals with early visual complaints are typically referred to eye specialists before referral to a neurologist. However, other problems with spelling, numeracy, and learned motor skills have also been reported. Even once an appropriate referral has been made, it may take some time before the diagnosis is made formally.

There is no diagnostic test for PCA. However, a number of tests may help to exclude potentially treatable causes such as infection or brain tumours. These may include some or all of the following:

- Specialised visual tests (by eye specialists)
- A full neuropsychological assessment (tests of thinking and reasoning abilities)
- Laboratory blood tests
- Brain imaging
- Lumbar puncture (examination of the fluid circulating around the brain and spinal cord)
- Other medical tests

The shrinkage of the back part of the brain as a result of brain cell loss may be visible on a brain scan. However, it may be difficult to diagnose, and a definitive diagnosis can only be made post-mortem once the brain tissue is examined by a pathologist.

The underlying cause of PCA in the majority of cases is Alzheimer's disease. However, other conditions can also result in similar initial symptoms. These include Lewy body disease, corticobasal degeneration and Creutzfeldt-Jakob disease. The exact prevalence of PCA in the population is currently unknown.

Progression of posterior cortical atrophy

As PCA progresses, word finding, day-to-day memory and general cognitive functions may become affected. In the later stages of the disease, people with PCA may develop jerking movements of their limbs and occasionally seizures. PCA causes progressive and irreversible decline in a person's skills and abilities over the years following diagnosis, and is a terminal disease.

Treatment of posterior cortical atrophy

Although there is no medication available to treat PCA specifically, patients may find some of the medications available for the treatment of typical Alzheimer's disease helpful. However, these medications are only designed to treat the symptoms of the disease. They may slow the disease progression but they are not a cure.

Although individuals with PCA often show relatively preserved insight, the cognitive problems associated with the condition often lead to a loss of independence, cause activities of daily living to be compromised, and inhibit the enjoyment of previous hobbies. Such problems can lead to depression, irritability, frustration and a loss of self-confidence. Individuals with PCA who are experiencing low mood may benefit from psychological therapies or antidepressant medication.

Practical visual aids designed to assist people with visual impairment may be of use to people with PCA. These include devices such as talking clocks or watches, mobile phones with simplified displays, cooking aids such as sensors that beep when a cup is almost full, and audio books.

Support for people with posterior cortical atrophy

Vision Australia provides advice and products that may be of help to people with PCA. Visit visionaustralia.org

The Royal Society of The Blind and Canberra Blind Society provide a full range of services for people who are blind or vision impaired. Visit rsb.org.au

The information in this help sheet is adapted with permission from information provided by the Dementia Research Centre, University College London, at raredementiasupport.org

Further Information

Dementia Australia offers support, information, education and counselling. Contact the National Dementia Helpline on **1800 100 500**, or visit our website at dementia.org.au



For language assistance phone the Translating and Interpreting Service on **131 450**