

Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative disorder that causes damage and deterioration to the back, or posterior, region of the brain. That area of the brain is responsible for processing what and how we see. PCA may affect a person's vision, their ability to read and write, their ability to navigate, and to reach for objects.

About posterior cortical atrophy

Posterior – the back part of the brain

Cortical – the thinking outer layer of the brain

Atrophy – shrinkage of that area of the brain

PCA is a rare form of dementia that is caused by the abnormal accumulation of amyloid plaques and neurofibrillary tangles in the brain. This build-up of plaques and tangles causes areas of the brain to become increasingly damaged and to shrink (atrophy) over time.

The part of the brain that is most affected by PCA is the posterior cortex, which is the back region (posterior) of the outer layer of the brain (the cortex). As the posterior cortex is responsible for processing visual information, PCA may significantly impair a person's visual functioning.

Since the presence of plaques and tangles in the brain is also a hallmark feature of Alzheimer's disease, PCA is referred to as a variant of Alzheimer's disease.

What are the symptoms?

Symptoms of PCA are most commonly seen in people in their 50s and 60s, but can affect people until their 80s. Early symptoms of this disease are often related to difficulties with a person's ability to process visual information.

Some of the common visual processing symptoms of PCA include: blurred vision, challenges with reading and writing, filling out forms, seeing objects in front of them, and problems with depth perception.

Other symptoms such as difficulties with simple math, knowing how to use tools and appliances, and knowing geographic directions may occur early in the disorder but unlike Alzheimer's disease, short-term memory is not a common symptom in the early stages of PCA. People in the early stages may experience increased anxiety, possibly because they are aware that something is wrong but have difficulties in explaining their symptoms.

Changes in cognitive abilities may be experienced as PCA progresses and causes more damage throughout the brain. This progression will often result in the onset of symptoms that are typical of Alzheimer's disease, like word finding and short-term memory loss.

How is posterior cortical atrophy diagnosed?

A diagnosis may take some time as people will often consult an eye doctor first because they think the symptoms they are experiencing are associated with their eyes. It can be challenging to diagnose PCA early in the progression of the disease. Many people are misdiagnosed because of their young age of onset and the unusual symptoms.

There is no single test to diagnose PCA but rather it is the recognition of the pattern of symptoms. Physicians assess the patterns and types of symptoms to help rule out other conditions that could be causing the symptoms, such as a brain tumor, a stroke or a treatable infection. A thorough assessment may also include testing of someone's thinking abilities, imaging of the brain with either a magnetic resonance imaging (MRI) or computerized tomography (CT), blood tests and specialized vision tests.

What are the causes or the risk factors?

Since the presence of plaques and tangles in the brain is characteristic of both Alzheimer's disease and PCA, research has found that in most cases of PCA, the underlying cause of the disease is Alzheimer's disease or another dementia.

Other neurodegenerative diseases, including: Lewy body dementia, Creutzfeldt-Jakob disease and corticobasal degeneration, also have been shown to cause PCA but this is very rare.

Is there treatment?

Currently, there is no known cure for PCA; however, some medications have been found to be effective in treating its symptoms. Cholinesterase inhibitors that are used to improve alertness and cognition in people with Alzheimer's disease can be used in persons with PCA.

Non-medication therapies can also be helpful including mental and physical exercises. Additional therapeutic approaches support people living with PCA to manage symptoms of their disease. Occupational and physical therapies can help to improve daily functioning. For example, an occupational therapist may provide guidance to people with PCA and their families to ensure that they can safely maneuver around their home, given their visual impairments. Practical visual aids, such as a talking clock, help to support the continued independence of people with PCA.

Support is available:

Visit the Alzheimer Society's website at www.alzheimer.ca or contact your local Alzheimer Society.

Additional resources:

- Alzheimer's Australia: http://www.fightdementia.org.au/common/files/NAT/20130912_NAT_HS_AboutDementiaHelpSheet_22.pdf
- Alzheimer's Society UK: http://www.alzheimers.org.uk/site/scripts/documents_info.php?documentID=135
- UCL Dementia Research Center: <http://www.ucl.ac.uk/drc/pcasupport>
- UHN Multidisciplinary Memory Clinic: Visual dysfunction in dementia, Home safety recommendations http://www.ucl.ac.uk/drc/pcasupport/contact_links/home_safety_tips

Two closed groups are available on Facebook. These groups are comprised primarily of PCA caregivers, and in some case the actual individual with PCA. Membership is available for people with PCA, their family, friends, colleagues, medical practitioners and support groups.

Contact the Group administrator for access. The groups are:

- 1) Posterior Cortical Atrophy-early stages
- 2) Posterior Cortical Atrophy Awareness

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