

Posterior cortical atrophy

Posterior cortical atrophy (also referred to as PCA and Benson's syndrome) is a rare progressive brain disease that is generally caused by the loss and dysfunction of brain cells, particularly at the back (**posterior**) part of the brain. As the disease progresses and more brain cells are lost, the brain tissue in the outer layer of the brain (known as the **cortical** area of the brain) shrinks (known as **atrophy**). The back part of the brain is responsible for processing visual information; therefore a person living with posterior cortical atrophy experiences **gradual vision decline** that does not get better with new glasses.

The loss of brain cells experienced by people living with posterior cortical atrophy is typically caused by the same brain changes seen in Alzheimer's disease.

A buildup of amyloid plaques and neurofibrillary tangles is believed to cause the loss of brain cells in people with posterior cortical atrophy. Because of similarities in the brain changes of Alzheimer's disease and posterior cortical atrophy, researchers believe that, in most cases, the underlying cause of posterior cortical atrophy is Alzheimer's disease.

Most people who have posterior cortical atrophy start noticing symptoms between the ages of 50 and 60, but it can also affect people older than 60.

Posterior cortical atrophy is diagnosed through the recognition of a pattern of symptoms, such as the slow onset of visual symptoms with no changes to memory and normal eye function.

A thorough assessment may include:

- Testing of cognitive functions such as perception, language and thinking abilities
- Imaging of the brain with either a magnetic resonance imaging (MRI) or position emission tomography (PET) scan
- Blood tests
- Specialized vision tests

An accurate diagnosis of posterior cortical atrophy may take some time. Because of the unusual combination of visual and cognitive symptoms, as well as the young age when symptoms commonly start appearing, many people are misdiagnosed with a different condition, such as a brain tumor, a stroke or a treatable infection.

Symptoms of posterior cortical atrophy

A person with posterior cortical atrophy may experience difficulties with:

- Reading and writing
- Interpreting what they are looking at
- Judging distances and depths – which may lead to car accidents or difficulties parking a car, and difficulties using stairs and escalators
- Orientation – a person may get lost, even in familiar settings, such as their home
- Recognition of objects and faces – especially when they are not right in front of the person
- Thinking skills – such as difficulties with simple math or spelling
- Using tools and appliances
- Blurry vision
- Sensitivity to light – such as glare from shiny surfaces
- Anxiety – which is especially noticeable in the early stages, possibly because the person is aware that something is wrong but has difficulties explaining their symptoms

As the brain damage progresses, changes in cognitive abilities, like difficulties finding words and short-term memory loss, may be experienced. These changes can be similar to those commonly seen in a person living with Alzheimer's disease.

The causes of posterior cortical atrophy are currently unknown.

Other neurodegenerative diseases, including [dementia with Lewy bodies](#) (sometimes referred to as Lewy body dementia), [Creutzfeldt-Jakob disease](#) and [corticobasal degeneration](#) have been shown to cause posterior cortical atrophy. However, this is very rare.

There are treatments that can manage the symptoms of posterior cortical atrophy.

- Occupational and physical therapies can help improve daily functioning. For example, an occupational therapist may provide guidance on how to move safely around the home.
- Practical visual aids and resources for people with sight problems, such as a talking clock and audiobooks, can support continued independence.

For information about medications that may be able to treat symptoms associated with posterior cortical atrophy, talk to your doctor or pharmacist.

There are many things that a person can do to live well with dementia, including making healthy food choices, being physically active, challenging the brain and participating in social activities. For brain-healthy choices and tips visit alzheimer.ca/brainhealth.

Support is available.

Contact your local Alzheimer Society for more information about dementia.

Visit alzheimer.ca/helpnearyou

Additional resources.

Visit brainxchange.ca and search “posterior cortical atrophy” to find webinars on this topic.

This resource is informed by research and the experiences of people living with dementia and their caregivers. We thank Dr. Isabelle Rouleau, Professor of Neuropsychology, Université du Québec à Montréal, for her generous contribution to the development of this resource.

To provide feedback on this factsheet, please email publications@alzheimer.ca