WHAT IS POSTERIOR CORTICAL ATROPHY?
INTRODUCTION

This booklet is for anyone who wants to know more about posterior cortical atrophy (PCA), an uncommon type of dementia. This includes people living with PCA, their family, friends and carers. We provide an overview of the causes, symptoms, diagnosis and treatments for PCA.

The information here does not replace any advice that doctors, pharmacists or nurses may give you. If you are worried about health, including issues with vision and memory, speak with your doctor as soon as possible.

This booklet was updated in August 2023 and is due to be reviewed in April 2025. It was produced in association with Rare Dementia Support, along with input from expert and lay reviewers. Please contact us if you would like a version with references or in a different format.

ANY QUESTIONS

If you have questions about dementia or dementia research you can contact the Dementia Research Infoline on 0300 111 5111 or email infoline@alzheimersresearchuk.org or write to us using the address on the back page.
WHAT IS DEMENTIA?

The word dementia is used to describe a group of symptoms. These include memory loss, confusion, vision and communication difficulties, and mood changes.

Dementia is caused by different diseases that affect the brain, such as Alzheimer’s disease. People living with dementia can have a wide range of symptoms, and these gradually get worse over time and affect day-to-day life.

WHAT IS POSTERIOR CORTICAL ATROPHY?

Posterior cortical atrophy (PCA) is an uncommon type of dementia that usually begins by affecting a person’s vision. It is also known as Benson’s syndrome, ‘visual variant’ or ‘visual-spatial’ Alzheimer’s disease.

PCA is caused by damage that builds up in the brain cells at the back of the brain. This is the part of our brain that processes information from our eyes, and allows us to make sense of what we are seeing and where things are. Alzheimer’s disease is most often the cause of the brain cell damage in PCA, but it is sometimes caused by other types of dementia, such as dementia with Lewy bodies.

Alzheimer’s usually affects a person’s memory first, but in PCA the first symptoms are often problems with vision and spatial awareness. Changes in someone’s ability to drive is often one of the first noticeable symptoms. It can also often cause difficulties with reading, locating, or recognising objects.

People who develop PCA tend to do so between the ages of 50 and 65, but it can affect older people too. PCA is a rare type of dementia, and at the moment we can’t be sure how many people around the world are affected by it. Around one in ten people diagnosed with Alzheimer’s disease at specialist dementia clinics might have symptoms of PCA.

POSTERIOR CORTICAL ATROPHY IS A RARE TYPE OF DEMENTIA WHICH OFTEN AFFECTS PEOPLE IN THEIR 50S AND 60S. IT CAN BE REFERRED TO AS YOUNG ONSET DEMENTIA.
SYMPTOMS

People living with PCA have symptoms that can vary from person to person and can change over time. Most people will have problems with their vision first, but some people may have problems with dressing, handwriting, coordination, numbers, and language too.

VISION

- Objects become less recognisable. People may struggle to recognise objects out of the corner of their eye or might see many different objects close together as one object.
- Surfaces and depth can look different. For example, a black object or puddle may appear to be a hole in the floor, or it may be difficult to find and reach for a door handle.
- People may not always be able to see what is right in front of them and may bump into things.
- Things may appear to have an unusual colour, appear distorted or look like they are moving around.
- People may still see an image of an object after looking away, or not be able to see more than one object at a time.
PEOPLE WITH PCA ALSO HAVE PROBLEMS WITH:

- **Reading:** Losing their place on a page, missing out lines, letters jumbling up or finding it hard to read certain fonts and handwriting. This can also affect everyday tasks like putting in a PIN on a cash machine.

- **Judging distances and depths:** Crossing roads and using escalators or stairs can become difficult. Moving around can also be difficult if there are lots of shadows, lights or patterns. A person may reach out to grasp an object but miss it.

- **Spatial awareness:** Some people may struggle with their sense of direction and terms like ‘left’ and ‘right’ may be harder to follow.

- **Recognition:** Problems recognising objects or faces, especially when they are not in plain sight. People may experience problems reading clocks or signs, especially digital clocks or screens.

- **Light sensitivity:** Finding bright light uncomfortable, including glare from shiny surfaces. People may have unusual colour experiences such as seeing patches of colour when it is dark.

- **Literacy:** Finding spelling and writing hard. People can find it difficult to remember the shape or name of certain letters.

- **Coordination:** Problems with dressing, for example, having difficulty using buttons or zips, locating the sleeves of a jacket while dressing, or putting clothes on back to front. Objects like kitchen utensils and remote controls may become difficult to use.

- **Numeracy:** Problems with simple calculations and dealing with money such as small change.

- **Mood:** Some people become low in mood, irritable or anxious, or may lose interest in things.

In PCA, the damage to brain cells spreads through the brain over time. This means that eventually a person’s memory, speech and problem-solving skills will be affected too.

AS SYMPTOMS PROGRESS, PEOPLE WILL NEED MORE SUPPORT IN THEIR DAILY LIFE AND HELP TO LOOK AFTER THEMSELVES. THIS CAN TAKE SEVERAL YEARS, BUT EACH PERSON’S PROGRESSION IS DIFFERENT AND UNIQUE.
DIAGNOSIS

Getting the right diagnosis is important so that people can get help and support. The symptoms of PCA can be very mild to begin with and difficult to describe. Because the condition is rare it can take some time to get the correct diagnosis. So, seeking help for symptoms as early as possible is recommended.

At first, people with PCA might think they have something wrong with their eyes and visit their optician. The optician may find that they have perfectly healthy eyes, or people could wrongly be given glasses that do not help them. This is because PCA damages the brain and not the eyes directly.

If you are worried about yourself or someone else who is showing signs of PCA, it is best to talk to your doctor. They will carry out tests to find out what is causing the symptoms.

These tests include:
- Questions about your symptoms and medical history.
- Speaking to your partner or someone close to you about the problems you are having.
- A physical check-up.
- A blood test to look for any underlying infections, vitamin deficiencies, thyroid disorders, or other conditions that could be causing the symptoms.

If your doctor suspects PCA or another form of dementia, they may send you to a specialist clinic for further tests.

These may include:
- Tests to work out how your brain processes the information it receives from your eyes.
- Memory and thinking skills tests.
- Brain scans to look for damage or loss of cells. This can be an MRI or CT scan and can also show if there are any signs of a stroke or brain tumour.
- A lumbar puncture that involves taking a sample of fluid from the bottom of your spine. This test can help to identify abnormal levels of proteins linked to diseases like Alzheimer’s in the brain.
- An EEG test, where sticky patches that measure electrical activity in the brain are placed on the head. This looks at how your brain cells communicate and does not hurt at all, the test takes about 20 minutes.

THE RESULTS OF ALL THESE TESTS WILL HELP A DOCTOR TO DECIDE THE LIKELY CAUSE OF YOUR SYMPTOMS AND DETERMINE WHAT TYPE OF DEMENTIA A PERSON MAY HAVE.
TREATMENTS

While there are currently no specific treatments that can stop or slow down PCA, there may be medicines that can help with the symptoms. For example, if Alzheimer’s disease is causing a person’s PCA they might be offered drugs called cholinesterase inhibitors.

These drugs work by helping brain cells to communicate with each other. They may help to improve a person’s symptoms for a time, but they do not stop the disease from getting worse. Some people find symptoms improve while taking these drugs, but others may not notice an effect.

There are three types of cholinesterase inhibitors:
- donepezil
- rivastigmine
- galantamine

Another drug called memantine may be prescribed for more severe symptoms, or if cholinesterase inhibitors haven’t worked.

Some people who have PCA caused by a form of dementia called dementia with Lewy bodies (DLB) may benefit from treatments that can help with movement problems. Levodopa is a drug used to treat both Parkinson’s disease and DLB, and people may also benefit from physiotherapy. Your doctor will be able to discuss these treatments with you if you have DLB and PCA.

Your doctor may also prescribe drug and non-drug treatments to help people with symptoms like depression and anxiety.

For more information, including some of the side-effects of these drugs, talk to your doctor or request our free ‘Treatments for dementia’ booklet.
WHAT AFFECTS MY RISK OF DEVELOPING PCA?

Researchers have found that the symptoms of PCA are caused by changes in the brain cells that process visual information from our eyes.

In PCA caused by Alzheimer’s disease, it is not clear why the disease affects this part of the brain more so than the areas affected by typical Alzheimer’s, which generally are involved in memory.

It is thought that what makes up a person’s risk of developing PCA is similar to other types of dementia. This is a complex mix of factors such as our age, the genes we inherit from our parents, and our environment and lifestyle. Scientists are working hard to find out more, as well as exploring specific genetic or lifestyle risk factors for the condition.
Some people with PCA may benefit from using visual aids and resources for people with sight problems. These may include audio books, devices with large and simple displays, voice recognition software and walking aids.

People may choose to carry a symbol cane to let others know they have sight problems.

For more information about visual aids and for support, contact the Royal National Institute of Blind People at helpline@rnib.org.uk or call 0303 123 9999.

People may also need to make changes to their home to help them stay independent and move around safely. There is some evidence that reading aids and home adaptations that support independent activities for people living with PCA can help.
HERE ARE SOME MORE WAYS TO HELP SOMEONE LIVING WITH PCA:

- At home, keep pathways clear by removing rugs, clutter and low furniture.
- Adding coloured stickers to glass doors can help someone see them more easily.
- Rooms should be well-lit. In the evenings or at night, nightlights may be helpful.
- Grab-bars in the bathroom and non-slip mats in the kitchen can be useful for safety and independence.
- Try to use plain furnishings, not patterned.
- Contrasting colours (for example, black and yellow) may help make objects clearer and mark edges, e.g. steps or kitchen worktops.
- At mealtimes, try to use plain plates and bowls, and cutlery with bright coloured handles may also help.
- Use a plain tablecloth and set out the plate, glass, etc. in the same way each time.
- Outdoors, different coloured paving may look like steps so try to let the person know that it is flat.

You can find out more about supporting someone with PCA by contacting Rare Dementia Support by emailing contact@raredementiasupport.org for details.

IT’S IMPORTANT TO REMEMBER THAT EVERY PERSON’S EXPERIENCE WITH PCA IS UNIQUE, AND WHAT WORKS FOR ONE PERSON MAY NOT WORK FOR ANOTHER.
SUPPORT FOR PCA

A diagnosis of PCA can leave a person with many questions. With the right information and support, people can live well and carry on doing the things they enjoy doing for some time. Talking to other people in the same situation can also help.

The Admiral Nurse Dementia Helpline offers practical and emotional support to anyone affected by dementia, including advice on managing the symptoms. Call 0800 888 6678.

The Rare Dementia PCA Support Group sends out newsletters and holds meetings across different parts of the country. These Support Group meetings provide opportunities for people affected by PCA to meet others and share their experiences. There is also a supportive Facebook group (search ‘Posterior cortical atrophy awareness’).

Visit raredementiasupport.org/posterior-cortical-atrophy/ for more information. You can contact them by email contact@raredementiasupport.org

The Alzheimer’s Society provides information and help for people with all forms of dementia and can tell you about local support groups in your area. Call 0333 150 3456.

You can also talk to your doctor or nurse for advice on caring for someone with PCA or ask for our booklet ‘Support for people affected by dementia: organisations that can help’.
RESEARCH

As the UK’s leading dementia research charity, we are working to revolutionise the way we treat, diagnose and prevent dementia. And then, we will find a way to cure it.

To do this, we’ve invested over £176 million in the best research, including £437,000 into PCA research, powering the most forward-thinking scientists and joining forces with world-class organisations.

Alzheimer’s Research UK have pioneered research into this condition as one of the first charities to invest consistently in this important area of research. For example, in 2013 we helped bring together international experts to form a working group to accelerate research into PCA. They compared people with PCA, typical Alzheimer’s disease and healthy individuals, and investigated changes over time. This research helped to improve our knowledge of how the disease spreads through the brain and what makes people vulnerable to different types of Alzheimer’s, like PCA.

With your support, we promise we will not stop until dementia can no longer destroy lives.

We are Alzheimer’s Research UK. We exist for a cure.

HOW TO GET INVOLVED

People with and without dementia, and dementia carers are needed for research studies.

If you’re interested in taking part in research and would like to find out more, you can contact Alzheimer’s Research UK’s Dementia Research Infoline on 0300 111 5111 or infoline@alzheimersresearchuk.org

You can register to the Join Dementia Research service, which is run by the NHS. This will match you to research studies you are suitable for, so you can see what type of research you could take part in.

YOU CAN FIND OUT MORE AND REGISTER HERE

joindementiaresearch.nihr.ac.uk You can also register over the telephone on 0300 111 5111
Alzheimer’s Research UK is the UK’s leading dementia research charity. We provide free dementia health information, like this booklet and others.

If you would like to view, download or order any of our other booklets please use the details below. If you’d like to help us review and improve our booklets, visit alzres.uk/reviewer

CONTACT US
0300 111 5111
infoline@alzheimersresearchuk.org

Alzheimer’s Research UK
3 Riverside, Granta Park, Cambridge CB21 6AD

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