WHAT IS PRIMARY PROGRESSIVE APHASIA?
INTRODUCTION

This booklet provides an overview of a condition called primary progressive aphasia (PPA), a rare form of dementia. It’s for anyone who is noticing symptoms in themselves or somebody else, or for people who want to know more about PPA and how it is diagnosed.

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

This booklet was updated in April 2023 and is due to be reviewed in April 2025. It was written by Alzheimer’s Research UK’s Information Services team in association with Rare Dementia Support, with input from expert and lay reviewers. Please get in touch using the contact details below if you’d like a version with references or in a different format.

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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 describes a group of symptoms that may include:
• memory loss
• confusion
• language and communication problems
• issues with sight
• mood and personality changes
• difficulty with day-to-day tasks.

The symptoms of dementia get worse as time goes on, so over time a person will need more help with everyday life. Dementia is caused by different diseases that affect the brain. These include Alzheimer’s disease, vascular dementia, dementia with Lewy bodies and frontotemporal dementia.

WHAT IS PRIMARY PROGRESSIVE APHASIA?

Primary progressive aphasia (PPA) is caused by diseases that affect parts of the brain that control our language, personality, emotions and behaviour. In most cases, PPA is caused by frontotemporal dementia. There are three separate types of PPA:
• semantic dementia
• progressive non-fluent aphasia
• logopenic aphasia.

MOST PEOPLE WHO DEVELOP PPA WILL BE IN THEIR 50s AND 60s.
SYMPTOMS

With all types of PPA, speech and language are affected. Symptoms will vary depending on what type of PPA someone has. However, in all types of PPA the speech and language difficulties progress and get worse over time.

SEMANTIC DEMENTIA

Over time, people with semantic dementia forget the meaning of words, as well as what objects and concepts are.

For example, they may:

- Have trouble using the right word – often saying another word or using a vague term like ‘thing’. As time passes, people will also struggle with more common words and often ask what words mean.
- Forget what items are used for. Such as every day or household items, tools and appliances.
- Talk about things at great length and in a vague or repetitive manner.
- Have difficulties understanding other people’s feelings.
- Find it difficult to focus on less familiar topics.
- Have problems reading and spelling.

In the later stages, they may:

- Tend to say less.
- Develop changes in behaviour and personality. Like sudden obsessions or mood swings, strong preferences for sweet or other specific foods, and difficulty understanding social situations.
- Find it difficult to recognise people they know, objects around the house or familiar sounds.
- Need much more assistance to manage day-to-day life.
PROGRESSIVE NON-FLUENT APHASIA

People with this condition have trouble speaking and make mistakes in how they say words or sentences.

For example, they may:
- Have trouble producing words, although they know what they want to say. Speaking may take a lot of effort and words may not come out right.
- They may speak more slowly, stutter and be hard to understand.
- Often confuse opposite words such as using yes when they mean no.
- Find that words come out in the wrong order or are missed out altogether.

Over time, they may develop other symptoms including:
- Problems with reading, writing and spelling.
- Difficulty hearing.
- Trouble understanding some words.
- Changes in behaviour and mood, for example becoming frustrated or depressed.
- Find it harder to make decisions or plans.
- Have difficulties swallowing food and drink.
- May develop similar symptoms to Parkinson’s disease such as slowness, stiffness or clumsiness of movements.
LOGOPENIC APHASIA

People with logopenic aphasia have trouble finding the words they want to use. For example, they may:

• Have trouble remembering the right word. The person may pause as they try to find the word they want.
• Speak more slowly and hesitantly and find it hard to say words correctly.
• Have trouble understanding and remembering more complex verbal information. This can make it hard to follow detailed conversations.
• Experience behavioural symptoms such as frustration, agitation and withdrawal from social situations.

As time passes, people can have more problems with their memory and thinking. For example:

• Forget conversations or appointments.
• Become lost and disorientated.
• Find it hard to do things like use the washing machine.

These later symptoms are similar to those in Alzheimer’s disease.

Someone with PPA may show symptoms of more than one type of PPA at the same time, or as their condition progresses. With all three types of PPA, problems get worse over time. It becomes harder for people to say what they need to, move about on their own and look after themselves.

Dementia is different for everyone who has it, and the speed at which it progresses can vary widely. Over time, someone with PPA will need more and more care and support until the end of their life.

YOU CAN SPEAK TO YOUR DOCTOR IF YOU ARE CONCERNED ABOUT ANY OF THE SYMPTOMS MENTIONED.
**DIAGNOSIS**

It is important to get a diagnosis of dementia as soon as possible, so that the right treatment and support can be given.

If you are worried about symptoms of dementia, your health or someone else’s, you should make an appointment with your doctor. After some initial tests, if your doctor suspects PPA or another form of dementia, they may send you to a specialist clinic.

**AT THE CLINIC**

Here, a doctor or nurse will run through more detailed tests with you. These may 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.
- Assessments of your thinking and language skills, memory, and changes to behaviour. Sometimes with a specialist speech and language therapist or psychologist.
- A brain scan, usually MRI.
- Possibly blood tests or other specialist tests such as a lumbar puncture.

Together, these will help a doctor to work out what is causing the speech and language difficulties and other symptoms.

PPA is a rare type of dementia, not often seen by doctors. This means it may take longer than usual to get the right diagnosis. When a diagnosis is made, many people will want to make plans for the future. This includes decisions about working, lifestyle and driving, care options, legal and money matters. Find out about help and support for families affected by dementia on page 18.

[alzheimersresearchuk.org](http://alzheimersresearchuk.org)
TREATMENTS

There are currently no medicines available to stop the progression of PPA, but there are treatments that may help with some of the symptoms.

A referral to a speech and language therapist can be useful at any time after a diagnosis. The therapist will assess your speech, language and communication difficulties and how they are affecting you or making everyday life difficult. They may also help with eating, drinking and swallowing difficulties.

People with PPA may also find it useful to see a physiotherapist, occupational therapist or dietitian for support with managing symptoms.

People with all forms of dementia may be offered antidepressants, or non-drug treatments like cognitive behavioural therapy (CBT). People with logopenic aphasia may be prescribed medicines commonly used to treat symptoms of Alzheimer’s.

For more information on these, ask for our booklet ‘Treatments for dementia’. You can also ask your doctor if these are suitable for you.
WHAT CAUSES PPA?

PPA is caused by diseases that affect brain cells in the front and sides of the brain, called the frontal and temporal lobes. Therefore, it is often referred to as a type of frontotemporal dementia. As with all types of dementia, over time brain cells become damaged and are lost. However, the causes for this loss in PPA are not yet clear.

Most cases of dementia are caused by a mixture of factors including age, genetics, lifestyle, and environment. We cannot change our age or genes. However, we can control some lifestyle factors that increase our risk of dementia.

Keeping socially connected, learning a new skill and regularly challenging your brain may help delay or reduce the impact of dementia. But they have currently not been shown to reduce the risk of PPA specifically. You can find more about how to reduce your risk of developing dementia by asking for our ‘Reducing your risk of dementia’ booklet.

In rare cases, semantic dementia or progressive non-fluent aphasia can be caused by a faulty gene that is passed down in families. The main genes involved are called MAPT, progranulin (or GRN) and C9ORF72. These genes are also associated with other forms of dementia. For more information about this, ask for our ‘Genes and dementia’ booklet.

We know that damage to brain cells in PPA is caused by an unusual build-up of certain proteins. This includes proteins called TDP-43 and tau, and researchers are working hard to find out why this happens and how it damages brain cells.

As frontotemporal dementia can be caused by the same proteins as motor neuron disease, symptoms of PPA can also often be seen in people with this condition. However, motor neuron disease symptoms only rarely develop in people with PPA.

Different types of PPA are often linked to different protein changes in the brain:

- **semantic dementia** is mainly linked to changes in the protein TDP-43,
- **progressive non-fluent aphasia** is most often linked to changes in the protein tau,
- most cases of **logopenic aphasia** are linked to the same underlying processes involved in Alzheimer’s disease. This includes the build-up of a protein called amyloid in the brain, which damages brain cells over time. This is why logopenic aphasia is often called an unusual or “atypical” form of young onset Alzheimer’s.

WE CAN CONTROL SOME LIFESTYLE FACTORS THAT INCREASE OUR RISK OF DEMENTIA.
HELP AND SUPPORT

A diagnosis of PPA will affect people in different ways and can come as a shock. People and their families may need time to come to terms with a diagnosis.

With the right information and support, people can carry on with their day to day lives for some time. Some people find that talking to others in the same situation can help.

The Young Dementia Network is a support group for people under 65 with any form of dementia, and their families. Their website, youngdementianetwork.org, has a range of resources, including information about local services. They also have a Facebook group: facebook.com/YoungDementiaNetwork

The national PPA Support Group holds several meetings a year in London, helping people living with PPA to meet and talk to others. Visit the PPA Support Group website at raredementiasupport.org/primary-progressive-aphasia, or email contact@raredementiasupport.org

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 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.

The PSP Association website, pspassociation.org.uk, has some helpful resources and an online forum. PSP (Progressive Supranuclear Palsy) is a type of dementia, with some symptoms that are similar to non-fluent PPA, including speech and swallowing problems.
SUPPORTING SOMEONE WITH PPA

For people with any type of PPA, their speech and language is affected first. When talking to someone with PPA, there are things you can do to help them:

• Be patient, warm and understanding.
• Find a quiet place to talk.
• Don’t ask more than one question at a time.
• Acknowledge their frustration when they show it, don’t dismiss it.
• Ask the person how you can help them communicate.
• Speak clearly and so that the person can see you speaking.
• Check that you have understood what they mean.
• One-to-one conversations can work better than talking in groups.

If someone is finding eating and swallowing hard, ask your GP for a referral to a speech and language therapist to assess their swallowing. This is really important, as there may be things they can suggest to reduce the risk of food and drinks going down the wrong way.

If someone is finding eating and swallowing hard, these may help:

• Try not to have foods that require a lot of chewing like thick pieces of meat.
• Cut food up into small, manageable pieces.
• Add sauces to meals.
• If eating becomes slower, a plate-warmer will help to keep food warm and nicer to eat.

Over time, someone with PPA will need more help with day-to-day life, for example with washing and dressing. It is important to think about safety at home and any changes you may need to make.

For people still working, PPA may make doing a job more difficult. It is helpful to talk to close family early on, about options such as lasting power of attorney, and later, home care and care homes.

Driving safety is also important. You should let the DVLA know if you or a family member is diagnosed with any form of dementia.

FOR ORGANISATIONS OFFERING HELP AND SUPPORT

Ask for a copy of our booklet ‘Support for people affected by dementia: organisations that can help’ or visit our website alzheimersresearchuk.org
RESEARCH

Alzheimer’s Research UK is the UK’s leading dementia research charity. We exist to change 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 dementia research, working with the most forward-thinking scientists and world-class organisations. This includes over £15 million of research into different types of frontotemporal dementia, including PPA. 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

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