



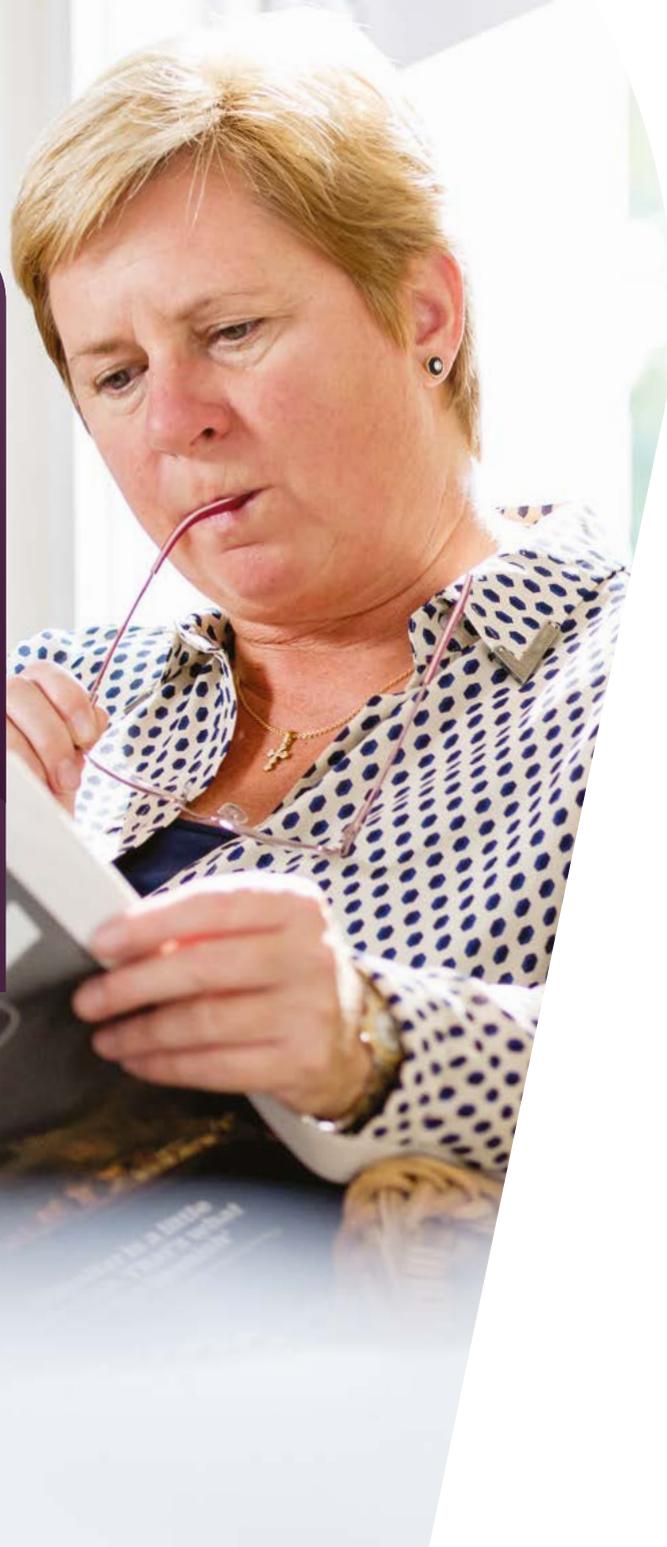
Alzheimer's
Research
UK

What is primary progressive aphasia?



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Introduction

This booklet is about a condition called **primary progressive aphasia (PPA)**. It is for people living with the condition, their family, friends, and carers. It provides an overview of the causes, symptoms, diagnosis, and treatments for PPA.

The information here does not replace any advice that doctors, pharmacists or nurses may give you. It provides background information that we hope you will find useful.

The booklet was updated in April 2021 and is due to be reviewed in April 2023. It was written by Alzheimer's Research UK's Information Services team and produced in partnership with Rare Dementia Support, with input from lay and expert reviewers. Please get in touch using the contact details below if you'd like a version with references or in a different format.

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



What is dementia?

The term dementia describes a group of symptoms that may include memory loss, confusion, language and communication problems, issues with sight, mood and personality changes and 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 diseases that affect the brain, there are many causes of dementia. 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 damage to parts of the brain that control our, language, personality, emotions and behaviour.

In most cases, this damage is caused by a group of diseases called frontotemporal dementia. Most people who develop PPA will be in their 50s and 60s.

The term PPA covers three separate conditions

- Semantic dementia
- Progressive non-fluent aphasia
- Logopenic aphasia



What causes PPA?

PPA is caused by damage to 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 dementia, over time brain cells become damaged and are lost, however, the causes for this loss in PPA are not yet clear. We know that there is an unusual build-up of certain proteins inside brain cells. These proteins include TDP-43 and tau, and researchers are working hard to find out why this happens and how it damages brain cells.

Different types of PPA often have different protein changes in the brain. Semantic dementia is mainly caused by protein TDP-43, while progressive non-fluent aphasia is most often caused by protein tau. Most cases of logopenic aphasia are caused by 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. Therefore, logopenic aphasia is often called an unusual or "atypical" form of early-onset Alzheimer's.

What causes a lot of dementia cases is a mixture of factors including age, lifestyle, environment, and genetics. While we cannot change our genetics, a lot of our risk can be modified by our lifestyle choices.



Keeping active and eating a healthy balanced diet, as well as looking after our heart health, will help to reduce our risk of developing PPA. 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 primary non-fluent aphasia can be caused by a faulty gene that is passed down in families. The genes involved are called MAPT, progranulin (or GRN) and C9ORF72. These genes are also associated with other forms of frontotemporal dementia. For more information about this, ask for our **'Genes and dementia'** booklet.

Symptoms

With all types of primary progressive aphasia, speech and language is affected. Symptoms will vary depending on what type someone has, but all symptoms 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'. This may start with names and other words they don't use very often. As time passes, people will also struggle with more common words and often ask what words mean.
- **Forget what every-day or household items**, tools and appliances are used for.
- **Talk about things at great length** and in a vague or repetitive manner.
- **Find it hard to understand what other people are saying.**
- **Have problems reading and spelling.**



In the later stages, people tend to say less. Changes in behaviour and personality are more common and occur earlier in semantic dementia than the other types of PPA. For example, people may develop obsessions or have mood swings, or act in ways that may seem strange to others.

Over time as the condition progresses they may find it difficult to recognise people they know, things around the house or familiar sounds. This means they need much more assistance to manage day-to-day life.

Progressive non-fluent aphasia

People with this condition have trouble producing speech 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.
- **Find that words come out in the wrong order** or are missed out altogether.
- **Stutter or speak more slowly** or hesitantly and be hard to understand.
- **Struggle to use the right grammar** when speaking or writing.
- **Find it hard to understand long and complex sentences.**

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 agitated or frustrated.
- **Find it harder to make decisions or plans.**
- **Trouble with swallowing.**
- **Similar symptoms to Parkinson's disease** such as shaking or being unsteady on their feet.

Logopenic aphasia

This form of PPA was identified more recently. People with logopenic aphasia are generally able to speak and understand others but 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.
- **Experience behavioural symptoms** such as frustration and agitation.

As time passes, people can have more problems with their memory and thinking. For example, someone may forget conversations or appointments, become lost, or struggle to use household appliances. These later symptoms are similar to those in Alzheimer's disease. You can speak to your doctor if you are concerned about any of the symptoms mentioned.

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 it progresses can vary widely. Over time someone with PPA will need more and more care and support until the end of their life.

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.



Tests at your doctors will 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 other aspects such as behaviour, sometimes with a specialist psychologist.
- You may also have blood tests, an MRI brain scan or other specialist tests.

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 14



Treatments

There are currently no medicines available to treat PPA, but there are treatments that can often help with some of the symptoms.

For example, your doctor can refer you to a Speech and Language Therapist. The therapist will assess your speech problems and how they are affecting you or making everyday life difficult.

They will help you find other ways to communicate if speech is difficult, including use of electronic devices. They can also help with eating, drinking and swallowing difficulties. People with all forms of dementia may be offered antidepressants, or non-drug treatments like cognitive behavioural therapy (CBT). You can ask your doctor if these are suitable.

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 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 www.raredementiasupport.org/primary-progressive-aphasia, email contact@raredementiasupport.org or call **020 3325 0828**.

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

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, www.pspassociation.org.uk, has some helpful resources and an online forum. PSP (Progressive Supranuclear Palsy) is a type of dementia, with some similarities to Parkinson's disease, but some of the symptoms are also similar to non-fluent PPA, including speech and swallowing problems.

You can also talk to your doctor or nurse for advice on caring for someone with PPA.

Supporting someone with PPA

For people with any type of PPA, their speech and language are affected first.

When talking to someone with PPA, there are things you can do to help them:

- Be patient and understanding.
- Find a quiet place to talk.
- Don't try to finish a person's words unless they ask you to help.
- Speak clearly and so that the person can see you speaking.
- Check that you have understood what they mean.
- One-to-one conversations generally work better than talking in groups.
- If the person starts to find talking hard, take a break so they can rest.
- They may find it easier to write things down or use an electronic aid than to talk.

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.
- If food is getting stuck in the throat, avoid dry and crumbly foods. Add sauces to meals and make sure people have plenty of water to drink with their food.
- 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.

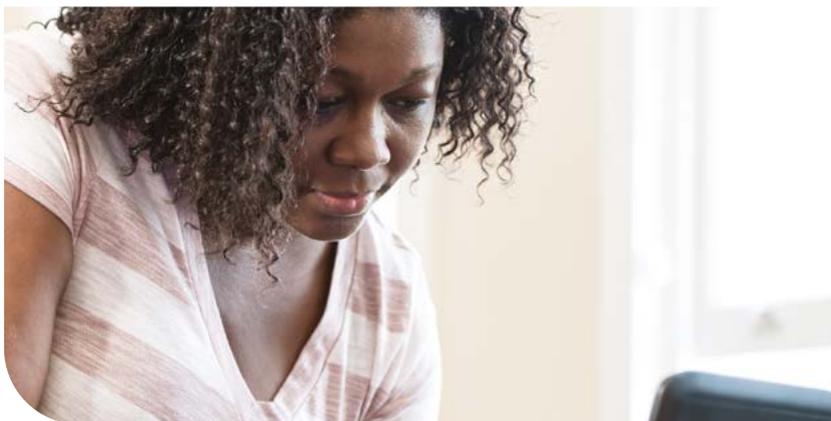
For organisations offering help and support, ask for a copy of our booklet '**Support for people affected by dementia: organisations that can help**' or visit our website www.alzheimersresearchuk.org

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 **www.joindementiaresearch.nihr.ac.uk** you can also register over the telephone on **0300 111 5111**.



Research

Alzheimer's Research UK has funded over **£12.6 million** of pioneering research to help advance our understanding of different types of frontotemporal dementia.

Our research is gaining new insights into the proteins that build up in the brain and how these damage nerve cells as well as taking these discoveries into drug discovery programmes to develop new treatments.

We have now funded over **£159 million** of pioneering research to help advance the understanding of all types of dementia, discovering new ways to diagnose, prevent and treat the diseases that cause dementia. Backed by our passionate scientists and supporters, we're challenging the way people think about dementia, and investing in research to make breakthroughs possible.

Alzheimer's Research UK is the UK's leading dementia research charity dedicated to making life-changing breakthroughs in diagnosis, prevention, treatment and cure.

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 details below.

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