

What is primary progressive aphasia?



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This booklet is about a condition called primary progressive aphasia (PPA). It's for anyone who may be worried about themselves or somebody else.

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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 helpful.

It was updated in April 2019 and will be reviewed in April 2021. Please get in touch if you'd like a version with references or in a different format.

This information booklet was produced in partnership with Rare Dementia Support.



The term dementia describes a group of symptoms including memory loss, confusion, language and communication problems, mood changes and difficulty with day-to-day tasks.

The symptoms of dementia get worse over time, so that a person will need more and more help with everyday life.

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 a condition caused by damage to parts of the brain that control our personality, emotions, language and behaviour.

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

The term PPA covers three separate conditions. In all three, people's speech and language are usually affected first, but in different ways:

Semantic dementia

Over time, people forget the meaning of words as well as objects and concepts.

Progressive non-fluent aphasia

People find it harder to speak and are more likely to make mistakes in how they say words or sentences.

Logopenic aphasia

People often pause while speaking as they try to find the right word.

Semantic dementia

People with semantic dementia gradually find it harder to remember the meaning of words. 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 everyday objects are for or find it hard to recognise them.

Talk about things at great length and in a vague or roundabout 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 in semantic dementia than the other types of PPA. For example, people may develop obsessions or a sweet tooth, or act in ways that may seem strange to others.

Later they may find it difficult to recognise people they know, things around the house or familiar sounds. This makes it harder to get on with day-to-day life.

Progressive non-fluent aphasia

This condition affects how a person produces speech. 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 letters or emails.

If you use sentences that are long and complex, someone with this form of dementia may find it hard to understand you. Over time, they may develop other symptoms including:

Problems with reading, writing and spelling.

Difficulty hearing.

Trouble understanding some words.

Changes in behaviour.

Trouble with swallowing.

Finding it harder to make decisions or plans.

Some people show signs similar to Parkinson’s disease such as shaking, being unsteady on their feet or having trouble using their hands. It’s also common for people to feel frustrated and low in mood.

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.

As time passes, people can have more problems with their memory and thinking. For example, someone may forget conversations or appointments, become lost, find it hard to work out how much change they need for a bus fare or struggle to use household appliances, like washing machines.

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 and move about on their own.

Dementia is different for everyone who has it, and the speed of change can vary widely. However, someone with PPA will need increasing care and support until the end of their life.



Diagnosis

It is important to get the right diagnosis so that the right help can be given. If you are worried about your health or someone else's, you should talk to your doctor.



If your doctor suspects PPA or another form of dementia, they may send you to a specialist 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, and memory.



You may also have blood tests, an MRI brain scan or other specialist tests.

Together all of these will help a doctor to work out what is causing the speech, language and other problems.

PPA is a rare condition, not often seen by doctors. 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 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 specifically for PPA, but there are treatments that might 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 and your family. They will help you find other ways to communicate if speech is difficult, including the 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). Ask your doctor for more information.



What causes PPA?

PPA is caused by loss of brain cells in the front and sides of the brain, called the frontal and temporal lobes. However, the reasons for this loss 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 but it's a complicated picture. Researchers are working hard to find out why this happens and how it damages brain cells.

Most people with logopenic aphasia have the same kind of damage in their brains as people with Alzheimer's disease. This includes the build-up of a protein called amyloid in the brain. Therefore, in most cases logopenic aphasia is an unusual form of early-onset Alzheimer's.

In a small number of people semantic dementia or progressive non-fluent aphasia may be caused by a faulty gene that can be 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, ask for our '**Genes and dementia**' booklet.

Help and support

A diagnosis of PPA will affect people in different ways, and they may need time to get used to it. With the right information and support, people can carry on with regular aspects of their lives for some time. Talking to other people in the same situation can help.

The **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/ppa, email contact@raredementiasupport.org or call **07388 220355**.

YoungDementia UK is a charity that offers support for people under 65 with any form of dementia, and their families. Their website, www.youngdementiauk.org, has a range of resources, including information about local services. They also have a Facebook group: www.facebook.com/YoungDementiaUK. Call **01865 794311**.

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**.

Alzheimer's Society provides information and help for people with all forms of dementia, and can tell you about local support groups and Dementia Cafés in your area. Call **0300 222 1122**.

The **PSP Association** website, www.pspassociation.org.uk, has some helpful resources and an online forum. PSP (progressive supranuclear palsy) is a different condition, but some of the symptoms are similar, including speech and swallowing problems, and occasionally PSP can begin with PPA.

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



Help and support

With all types of PPA, speech or language is 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 loudly and clearly.
- Check that you have understood what they mean.
- One-to-one conversations may 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 ideas may help:

- Try to avoid thick pieces of meat, or cook them slowly so they are easier to chew and swallow.
- 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.



With time, someone with PPA will need more help with day-to-day life. It is important to think about safety at home and any changes you may need to make. In those of working age, PPA may make working life 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



Research

Alzheimer's Research UK has funded over

£21.9 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.

Backed by our passionate scientists and supporters, we're challenging the way people think about dementia, and investing in research to make breakthroughs possible.



Find out more

If you have questions about dementia research or want to find out more about how to get involved in research, contact our **Dementia Research Infoline** on **0300 111 5 111** or email infoline@alzheimersresearchuk.org

The Infoline operates 9.00-5.00pm Monday to Friday. Calls cost no more than national rate calls to 01 or 02 numbers and should be included in any free call packages.

Freeport RTYR-ZUUZ-AULL
Partridges
3 Eagle Avenue
Magnetic Park
Desborough
NN14 2WD



Send me more information

For free information, simply complete this slip and drop it straight in a post box. Alternatively, phone us on **0300 111 5555**.

I would like to know more about

Dementia: symptoms, diagnosis, causes, and risk reduction (SCIHIAAD)

Treatments for dementia (SCIHITMT)

Genes and dementia (SCIHIGENE)

Support for people affected by dementia: organisations that can help (SCIHCARE)

The latest dementia research (SMTTHINK)

Name

Address

Email

We'd like you to be the first to know about the latest research and how your support makes a difference, as well as ways you can get involved and help fund our life-changing work. We'll keep your information safe and never sell or swap it with anyone.

Let us know how we can contact you (tick below):

Post Email Telephone Text message

You can change how we talk to you at any time, by calling **0300 111 5555** or emailing enquiries@alzheimersresearchuk.org

Our Privacy Notice can be found at www.alzheimersresearchuk.org/privacy-policy and explains how we will use and store your information.

PPA19

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

We welcome your comments to help us produce the best information for you. You can let us know what you think about this booklet by contacting us using the details below.



Contact us

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