

Who looks after people with PID?

Doctors and nurses who are specialists in immunology (the working of the immune system) look after people with a PID. Specialists are needed because PIDs are rare, they can cause severe complications and complex treatments are sometimes needed.

A consultant immunologist will lead your care, and your main point of contact with your health team is likely to be via an immunology clinical nurse specialist. It is extremely important that those affected receive expert care and advice from a consultant immunologist at one of the specialist centres in hospitals throughout the UK.

Your care team may also involve other specialists, including doctors who are experts in other branches of medicine, e.g. respiratory (lung) and gastroenterology (gut) physicians.

What treatments are available?

The treatment you receive will be dependent on the type of PID you have and how it affects your immune system. A large proportion of people with a PID have immunoglobulin replacement therapy to help keep them free from infection and are given antibiotics as and when an infection occurs. Some other types of PID may involve taking antibiotic and/or antifungal medicine daily to stave off infection. More specialised treatments and potential cures for some types of PID include bone marrow transplantation, enzyme replacement therapy and gene therapy. Research is leading to a greater understanding of the causes of PID, leading to better-tailored treatments for those affected.

Taking preventative measures to avoid exposure to infection also plays a key role in keeping people with a PID well. Please see our leaflet on 'Keeping well and healthy when you have a PID'.

How will my health be monitored?

Doctors will monitor your health by clinical review (check-up) and infrequent blood tests. Your GP will be involved in collaboration with the specialist centre.

About Primary Immunodeficiency UK

Primary Immunodeficiency UK (PID UK) is a national organisation supporting individuals and families affected by primary immunodeficiencies (PIDs).

We are the UK national member of the International Patient Organisation for Primary Immunodeficiencies (IPOPI), an association of national patient organisations dedicated to improving awareness, access to early diagnosis and optimal treatments for PID patients worldwide.

Our website at www.piduk.org provides useful information on a range of conditions and topics, and explains the work we do to ensure the voice of PID patients is heard.

If we can be of any help, please contact us at hello@piduk.org or on **0800 987 8986**, where you can leave a message. Visit www.piduk.org for further information.

Support us by becoming a member of PID UK. It's free and easy to do via our website at www.piduk.org/register or just get in touch with us. Members get monthly bulletins.

PID UK is reliant on voluntary donations. To make a donation, please go to www.piduk.org/donate



Primary immunodeficiency - the basics

hello@piduk.org
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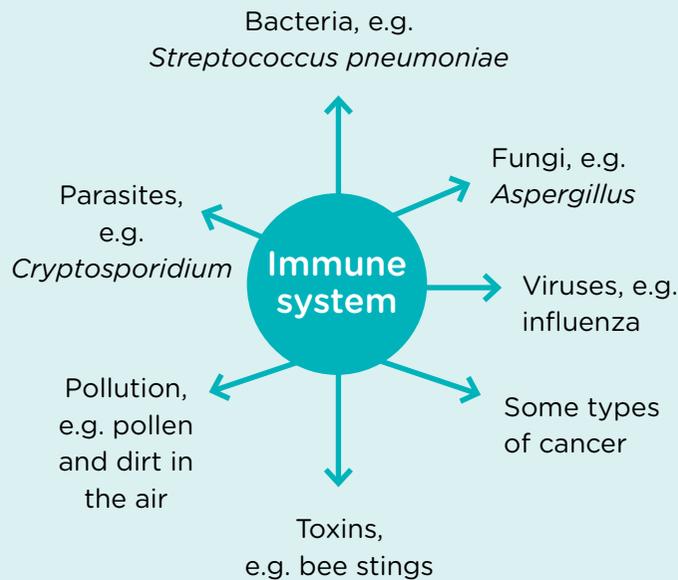
This leaflet helps answer some of the questions patients, parents and carers may have about primary immunodeficiency (PID) when a diagnosis has been made.

Primary immunodeficiency, often shortened to PID, is a term used to cover a large number of different conditions that affect how the body's immune system works. Sometimes these conditions are also referred to as primary immune deficiencies.

What is the immune system?

The immune system is made up of specialised cells, proteins, tissues and organs that defend people against germs (micro-organisms), and some types of cancer. It works to keep people healthy and to prevent infection. When problems with the immune system occur, it can lead to illness and infection.

The immune system helps protect against a wide range of harmful agents.



What happens in PID?

People with a PID have parts of their immune system missing or not working correctly, hence the term 'deficiency'. This leaves them with reduced or no natural defence against germs, such as bacteria, fungi and viruses, which surround us every day.

The consequences are that people with PID get infections more frequently than is usual and the infections can take longer to get better. When antibiotic treatment is required, PID patients are often prescribed longer and stronger courses. Even then sometimes the infections can come back.

Susceptibility to infection is one of the most common symptoms of PID. Often PID is diagnosed early in a child's life but signs of immunodeficiency can also occur for the first time in older children, teenagers or adults.

Occasionally other medical problems can arise as complications of PIDs. These include 'autoimmune' and 'inflammatory' problems, where the immune system reacts inappropriately against the body's own tissues, as well as a slightly increased risk of cancer in some conditions.

How do people get PID?

PIDs are mainly genetic disorders, meaning they are inherited and can be passed on from one generation to the next. They are caused by errors in the genes and proteins of the cells that make up the immune system. Most people with PID are born with the condition.

In PID some of the DNA building blocks that make up the genes involved in the workings

of the immune system are either not there or altered ('mutated'). This results in either missing or faulty proteins, causing essential parts of the body's defence against infection not to be made or work properly. For many PIDs the precise genetic mistake is now known, and understanding of these conditions is increasing all the time.

However, in some types of PID (e.g. common variable immune deficiency (CVID)), the link to specific faulty genes is not yet well understood and it is less common for parents to pass on the disorder to their child. Progress is being made but it may be a long time until we find all the causes. This may change as technology for the screening of genes improves and more research is done.

You certainly cannot catch a PID from those affected, and they cannot transfer their disorder to anyone else through contact.

Over 300 different PID conditions involving different parts of the immune system have now been identified and about 5,000 people have a PID in the UK.

Why are they called primary?

They are called primary because of the basic, built-in nature of the immune deficiency. The term is used to separate PID from conditions where the immune system is affected by secondary causes, such as other diseases (e.g. HIV), drug treatment, chemotherapy, malnutrition or environmental exposure to toxins.

There is no connection to HIV/AIDS, which is a secondary or 'acquired' immunodeficiency.