

What is the main treatment for primary antibody deficiency?

Milder antibody deficiencies may require prompt treatment at the earliest sign of infection or the use of preventative antibiotics. Immunoglobulin therapy is offered to people affected by more severe primary antibody deficiency. Therapeutic immunoglobulin products contain a wide mixture of antibodies that help fight infections. Immunoglobulin is made from screened, donated, blood-derived plasma. During manufacture everything except IgG is removed from the plasma. IgG is very good at fighting bacteria and viruses.

Immunoglobulin therapy can be given through a vein or through the skin but must be given regularly. Although immunoglobulin treatment is very effective, infections can still occur, and these require prompt treatment with antibiotics.

More information about immunoglobulin therapy can be found in our patient booklet 'Immunoglobulin therapy'.

Other PID UK leaflets that may be useful to people affected by PAD include:

- Keeping well and healthy when you have a PID
- Making the most of your appointments
- Going on holiday
- Insurance matters
- Your employment rights
- Looking after your lungs
- Antibiotics in PID

Please get in touch with us to request copies.

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 and newsletters twice a year.



PID UK is part of Genetic Disorders UK (registered charity number 1141583 and registered company number 7564771). The registered office is at 199A Victoria Street, London SW1E 5NE.

Primary antibody deficiency



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This leaflet is for people who have been diagnosed with a primary antibody deficiency and should be read in conjunction with our leaflet on the basics of primary immunodeficiency.

What is primary antibody deficiency?

Primary antibody deficiency, or PAD for short, is the name given to a number of different conditions that are part of a larger group of disorders known as primary immunodeficiencies (PIDs).

The most common types of primary antibody deficiency include common variable immune deficiency (CVID), X-linked agammaglobulinemia (XLA), hyper IgM syndromes, selective IgA deficiency, IgG subclass deficiency and specific antibody deficiency. You can access our condition-specific booklets on the PID UK website at www.piduk.org/resourcesforpatientscarersandprofessionals.

Some people have low levels of antibodies as a result of previous treatment or owing to other medical problems. In these cases the antibody deficiency is referred to as secondary antibody deficiency.

Why does primary antibody deficiency occur?

Primary antibody deficiency results from the failure of the immune system to produce sufficient antibodies to fight infections, particularly those caused by bacteria.

The underlying problem causing the inability to make sufficient antibodies is not the same in every person who has an antibody deficiency as there are multiple steps and pathways for the production of these important defenses, any one or more of which may be defective.

What are antibodies?

Antibodies are members of a family of proteins called immunoglobulins (Ig). They work to fight off infections. They are produced by specialist white blood cells and are present in body fluids.

Why are antibodies important?

Antibodies do two things:

- They help us to recover from an infection.
- They help to prevent the same infection from coming back in the future.

Antibodies are divided into five main classes or kinds of immunoglobulin. Each class of antibody has a different function.

IgG is the most abundant and common immunoglobulin, found in blood and tissue fluids. IgG functions mainly against bacteria and some viruses.

IgA is found in nasal fluids, bile, tears, sweat and saliva. It protects the tissues of the respiratory (lungs), reproductive, urinary and digestive systems.

IgM is a rapid response antibody and is the first type of protective antibody produced in response to infection.

IgE is responsible for allergic reactions; e.g. against pollen, causing hay fever.

IgD helps activate the cells that make antibodies and other types of immune cells, causing them to release antimicrobial chemicals. It is not routinely measured.

All types of immunoglobulin are made up of antibodies against the germs that an individual has encountered during the course of his or her life.

Antibodies normally get into all parts of the body, including the skin, eyes, ears, nose, throat, lungs, intestines and joints to fight against invading organisms. Consequently any part of the body can be infected when antibodies are missing, as in primary antibody deficiency, although usually only a few parts are affected as the germs are particular about where they invade.

Which cells make antibodies?

Antibodies are produced by specialist white blood cells (plasma cells / B-cells). Plasma cells are a specific subtype of B-cell found within the bone marrow or lymph nodes. They are responsible for the majority of high-quality antibody production. The long life of plasma cells helps people with healthy immune systems to retain immunity to viruses and bacteria that infected them many years ago or to which a person was immunised.

Doctors are able to identify what type of primary antibody deficiency a patient has by measuring the levels of immunoglobulin, and the number and function of B-cells.

How do antibodies protect against infections?

Antibodies help the body fight infection in a number of different ways. For example, antibodies can bind to the surface of a virus and interfere with the virus's ability to attach to cells in the body. Antibodies can also attach to the surface of some bacteria and viruses, causing activation of other parts of the immune system that can directly kill these microorganisms. Antibody-coated bacteria are also much easier targets for other white blood cells to ingest and kill than bacteria that are not coated with antibodies.

All these actions of antibodies prevent microorganisms from successfully invading body tissues and causing serious infections.

