

Stay healthy! A guide for patients and their families



Primary immunodeficiencies — Stay healthy! A guide for patients and their families
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What are primary immunodeficiencies?

This booklet explains what primary immunodeficiencies (PIDs) are and what steps people with PIDs should take to stay healthy.

PIDs are a large group of disorders caused when some components of the immune system (mainly cells and proteins) do not work properly. Some PIDs are relatively mild, while others are severe. PIDs are often identified during childhood, but they can also be diagnosed in adults. PIDs are not related to AIDS ('acquired immunodeficiency syndrome'), which is caused by a viral infection (HIV).

The immune system normally helps the body fight off infections by germs (or 'micro-organisms') such as bacteria, viruses, fungi and protozoa. As their immune systems do not work properly, people with PIDs are more prone than other people to infections. These infections may be more common than is usual, they may be particularly severe or difficult to clear, or they may be caused by unusual micro-organisms. They may occur whatever the season, even in summer.

Many people with PIDs receive treatment with immunoglobulin replacement, which helps to protect against infections. Other treatments for PIDs include stem cell (or bone marrow) transplantation for some patients with certain severe PIDs, granulocyte-colony stimulating factor and gamma interferon. The IPOPI booklet '*Primary immunodeficiencies — Treatments for primary immunodeficiencies: a guide for patients and their families*' explains more about these treatments.

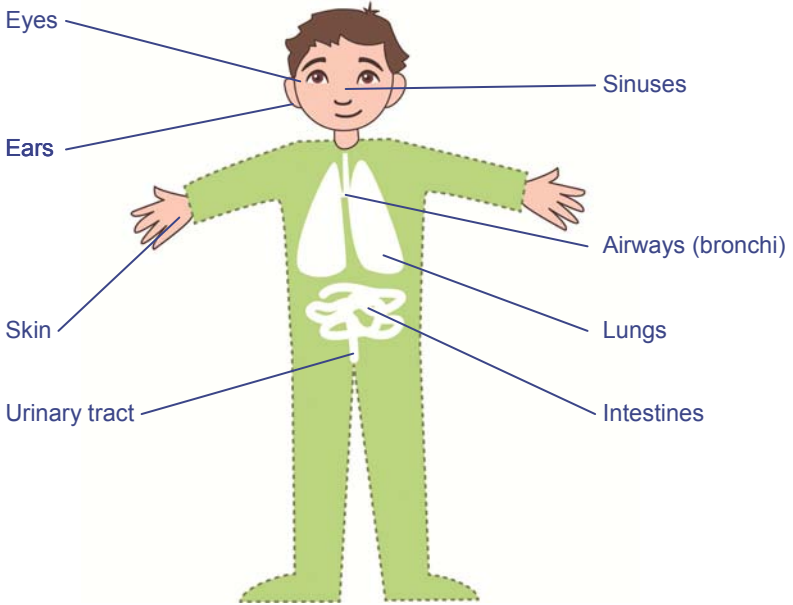
PIDs are caused by hereditary or genetic defects of the immune system. PIDs are not related to AIDS ('acquired immunodeficiency syndrome'). PIDs are not contagious — it is not possible to 'catch' a PID or to spread it to other people. However, children can inherit PIDs from their parents. People with PIDs should therefore seek advice on the genetics of their condition if they wish to have children.

PIDs can also cause the immune system to attack the body itself — this is called 'auto-immunity'. This can cause pain and swelling in the joints, known as 'arthritis'. It can also cause skin rashes, a loss of red blood cells (anaemia) or platelet cells involved in blood clotting, inflammation of blood vessels, diarrhoea and kidney disease. Patients with some PIDs are also more likely to have allergies and asthma.

Treatments for PIDs allow many patients to enjoy lives that are as full and normal as possible. There are lots of things that adult patients, parents of children with PIDs and children themselves can do to stay healthy.

Preventing infections

People with PIDs are prone to infections in various parts of the body, some of which are shown below. It is important to contact the doctor whenever an infection is suspected.



Most importantly, there are various ways that patients and parents can help prevent infections.

Hygiene

Good hygiene is very important. This means simple things, such as:

- Washing the hands regularly and carefully, especially before meals and after using the toilet, outdoor activities and playing with pets.
- Cleaning and dressing cuts and scrapes.
- Good personal hygiene, which is very important, including careful brushing of the teeth and regular visits to the dentist.
- Good food hygiene, to help avoid food poisoning.

Everyone should cover the mouth and nose when sneezing and coughing.

- Good home hygiene — it is important to keep the house clean and to avoid high humidity levels to prevent lung problems. Children's toys should also be cleaned regularly!
- Avoiding smoky and crowded places.

Where possible, people with PIDs should try to avoid contact with people with infections. For example, parents of children with severe PIDs should ask the school to inform them of any infection outbreaks and then ask for advice from the immunology team.

Some people with PIDs may need to take precautions when having contact with pets and other animals. In case of doubt, ask for your doctor's advice.

If surgery is needed it is important that the surgeon knows that a person has a PID so that steps are taken to prevent infections related to the surgery.

Vaccination

Vaccination (or 'immunization') involves giving a small dose of bacteria or virus that causes an infection in order to help the body become immune to it. Some vaccines contain killed micro-organisms, while others contain live micro-organisms.

Most patients treated with immunoglobulin replacement therapy generally do not need vaccines. Importantly, most patients should not be given 'live-attenuated' vaccines, as these can cause them to have infections. Live-attenuated vaccines include rotavirus, oral polio, measles, mumps and rubella (MMR[®]) and varicella vaccines, and the 'BCG' vaccine for tuberculosis. The family of a patient should normally be vaccinated in order to keep infections from the home. However, patients and parents should ask their doctor's advice in all matters regarding vaccination.

Many patients receive immunoglobulin replacement therapy to provide normal levels of antibodies to protect them against infections. People with PIDs also often require medicines to treat or prevent infections caused by bacteria (antibiotics), viruses (antivirals) or fungi (antifungals). As with all prescribed medicines, it is important to follow the instructions given by the doctor, nurse or pharmacist.

Travel

Having a PID should not prevent patients travelling, but certain precautions are advisable. Patients should discuss travel plans with their immunologist, who can advise on safety issues and on the need for vaccines. Before travelling, people with PID should ensure they have adequate medical and travel insurance. If possible, they should ask their doctor or specialist centre to arrange any scheduled therapy needed during their travels.

Patients should carry a statement explaining their condition and the purpose of the drugs and other equipment carried. This can be in a letter signed by their immunologist and if possible translated into the language of the countries to be visited. Sample letters can be found at www.ipopi.org.

Further help in each country can be provided by national PID patient organisations and in some specific instances by the local blood bank or Red Cross Blood Transfusion Service.

Diet

A nutritious, balanced diet is important to everyone's health. People with PIDs should avoid raw or undercooked dishes (e.g. meats, eggs and cheeses). Most do not need a special diet or supplements such as extra vitamins. Patients and parents should consult the immunology team before any supplements are taken.

People with PIDs should avoid drinking water of unknown origin, or water that has remained too long in the same container.

Skin care

Some people with PIDs may need to pay particular attention to skin care, such as precautions against sun exposure. In case of doubt ask for your doctor's advice.

Exercise and sports

People with PIDs can enjoy exercising and sports, just like everyone else. Patients with phagocyte cell deficiencies (such as chronic granulomatous disease, or CGD) should avoid swimming in lakes or ponds, and gardening that involves digging or contact with rotting plant or tree material. Certain PIDs interfere with blood clotting and patients with these conditions need to discuss safe sports with their immunologist.

Sleep

People with PIDs should make sure they get plenty of sleep, as this is important to health. The right amount of sleep will depend on the patient's age. If they are in any doubt, parents and carers should ask their doctor or nurse how many hours of sleep their child needs.

Sexual life

People with PIDs should take precautions to avoid sexually transmitted diseases, for example through the use of condoms.

People with PIDs should not smoke, as this can cause chest infections. If possible, they should ask others around them not to smoke too. Parents of children with PIDs should not smoke.



Primary immunodeficiencies

Further information and support

This booklet has been produced by the International Patient Organisation for Primary Immunodeficiencies (IPOPI). A companion booklet titled '*Primary immunodeficiencies — Treatments for primary immunodeficiencies: a guide for patients and their families*' is also available.

For further information, and details of PID patient organisations in 40 countries worldwide, please visit **www.ipopi.org**.

Provided by



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