Secondary immunodeficiency (SID)

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PID UK
Supporting families affected by primary immunodeficiencies

About this booklet

This booklet provides information on secondary immunodeficiency. It has been produced by the PID UK Medical Advisory Panel and reviewed by patients affected by secondary immunodeficiency to help answer the questions other patients and their families may have about this condition. The information should not, however, replace advice from a medical professional.

Secondary Immunodeficiency (SID)
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What is secondary immunodeficiency?

Secondary immunodeficiency (SID) occurs when the immune system is weakened by another treatment or illness.

What causes secondary immunodeficiency?

There are many potential causes of SID but the most common examples are blood or bone marrow disorders, drugs (medicines) and treatment for cancer. Some cancers can be responsible for SID, too.

The main medicines that cause problems are those that target the immune system, including immunosuppressant drugs, 'biologics' and chemotherapy. These medicines may have been used in the treatment of conditions such as rheumatoid arthritis, multiple sclerosis, inflammatory bowel disease and psoriasis, and of blood and lymph node cancers in particular.

There are other medicines that are recognised as causing specific complications with the immune system and these effects are not directly related to the way the drug works. For example, some anti-epileptic drugs can cause antibody deficiency.

There are some rare, inherited medical problems, such as transcobalamin deficiency and gut lymphangiectasia, that are not immune deficiencies but cause failure of antibody production or loss of antibodies from the gut. These causes can be treated and/or replacement immunoglobulin therapy given.

Although HIV infection is a cause of SID, this is usually treated as a separate problem in its own right. However, it is often something that doctors wish to exclude before considering other causes for primary or secondary immunodeficiency.

How will I know if I have a secondary immunodeficiency?

The signs and symptoms of SID are the same as for primary immunodeficiency; that is, frequent, prolonged or unusual infections. Tests are usually necessary to understand what the problem may be.

What tests will be performed?

Blood tests are used to test for SID. The precise tests of immune function will depend on the type of infections that have been a problem. It may be that you have low numbers of neutrophils. This can easily be detected by repeating a full blood count on several occasions. Or, it may be that you aren't making enough antibodies. Other, more subtle, immune defects may require blood tests that are a little more complicated from a laboratory point of view, however, these will only require a few blood samples from you.

What treatment is available?

The treatment you will be recommended depends on the nature of the deficiency. If an ongoing treatment is thought to be the cause, then the treatment may be changed or removed. If another disease is the cause, then treating it may resolve the immunodeficiency.

Antibody deficiency
SID may mimic any of the primary antibody deficiencies (see the leaflet at **www.piduk.org/static/media/up/primaryantibodydeficiency.pdf**), and the level of treatment depends on the severity. It may be that a test vaccination is recommended which, if successful, will stimulate your body to make good protective antibodies; in which case, no additional therapy is needed. Some patients may require preventative (prophylactic) antibiotics. A few people may need to have immunoglobulin replacement therapy (see **www.piduk.org/static/media/up/PIDUKImmunoglobulin.pdf**).

Neutropenia
Low neutrophil counts can be caused by immunosuppression or other medical treatments and are an indication for your doctors to change your therapy. If this is not possible or the treatment has already stopped, patients may require both antibiotic and anti-fungal prophylaxis. Sometimes an injectable treatment (G-CSF; granulocyte – colony stimulating factor) is used that stimulates the bone marrow to make and release more neutrophils.

T-cell deficiency
Defects in T-cell function or number may be very clinically significant and the health problems caused may get better if the causative medicine can be removed. Patients with poor T-cell number or function may often suffer from infections with viruses, bacteria and 'atypical' organisms such as the usually less troublesome members of the tuberculosis (TB) family – none of which causes problems in a healthy individual.

If the underlying cause cannot be removed or resolved, then the treatment is supportive, with anti-viral drugs (where appropriate), anti-fungals and/or antibiotics.

For individuals who have poor T-cell function as a consequence of a bone marrow transplant for cancer, it may be possible to 'top up' their immunity with an additional treatment of donor lymphocytes (donor lymphocyte infusion; DLI). For some patients the treatment for graft-vs-host disease may have resulted in the immune deficiency, and in such cases a DLI is usually not possible.

Will my treatment be life-long?

If the underlying cause can be removed, then in many individuals the immune system returns to normal. For others, this is not the case and the treatment is long term, even life-long.

How will I know if I don't need treatment anymore?

Your immune system will be regularly monitored to determine if you need continuing treatment.

Can I pass on or give this problem to someone else?

These problems are not 'infectious', unless the problem is HIV.

Glossary of terms

antibody a type of protein (immunoglobulin) that is produced by certain types of white blood cells. Antibodies fight bacteria, viruses, toxins and other substances foreign to the body.

biologics drugs that are genetically engineered proteins derived from human genes. They are designed to inhibit specific components of the immune system that play important roles in driving inflammation.

bone marrow spongy tissue found in the hollow centres of some bones. It contains stem cells that can grow into any of our normal blood cells.

bone marrow transplant (BMT) transfer of bone marrow, obtained usually from the hip bones, from a healthy donor – either related or unrelated – to another person, e.g. a patient with cancer or a primary immunodeficiency. The donor bone marrow replaces the patient's bone marrow and provides a new immune system. Sometimes this is referred to as stem cell transplantation.

chemotherapy treatment with cytotoxic drugs that kill cells in the body. In cancer, chemotherapy is used to kill cancer cells. In stem cell transplantation, chemotherapy is used to suppress the immune system so that the patient will not reject the new bone marrow and to make room in the bone marrow for the donor marrow stem cells to grow.

deficiency a lack or shortage.

donor lymphocyte infusion (DLI) a procedure in which white blood cells are collected from a patient's stem cell donor and infused into the patient sometime after transplantation.

epileptic/epilepsy a condition that affects the brain. When someone has epilepsy it means they have a tendency to have epileptic seizures.

granulocyte – colony stimulating factor (G-CSF) a type of growth factor. Growth factors are proteins made in the body and some of them make the bone marrow produce blood cells. G-CSF makes the body produce white blood cells to reduce the risk of infection after some types of cancer treatment. It also makes some stem cells move from the bone marrow into the blood.

gut lymphangiectasia dilatation of gut lymphatic vessels and loss of lymph fluid into the gastrointestinal tract, with associated loss of proteins, including immunoglobulins.

HIV human immunodeficiency virus, causing the acquired immunodeficiency syndrome (AIDS).

immune system the structures and processes that protect the body against infection and disease.

immunodeficiency when the immune system's ability to fight infectious disease is compromised or entirely absent.

immunoglobulin replacement therapy a plasma-based treatment. The immunoglobulin contains antibodies that help fight infection. This treatment can be given through a vein or through the skin.

immunosuppressant a type of drug that suppresses or reduces the strength of the body's immune system.

inflammatory bowel disease (IBD) the name of a group of disorders in which the intestines (small and large intestines or bowels) become inflamed (red and swollen).

lymph nodes small, bean-sized organs of the immune system distributed widely throughout the body. They are the home for the many types of cells that are important in fighting infections.

lymphocyte a white blood cell that works to fight infection in the body. One type of lymphocyte is called a 'B-cell'. This type of lymphocyte makes antibodies.

multiple sclerosis a disease in which the insulating covers of nerve cells in the brain and spinal cord are damaged.

neutropenia a low level of neutrophils, a type of white blood cell.

neutrophil a type of white blood cell.

primary antibody deficiency covers a range of disorders resulting from the failure of the immune system to produce sufficient antibodies in the bloodstream to fight infections.

prophylactic something that works to defend or protect against disease.

psoriasis a skin condition that causes red, flaky, crusty patches of skin covered with silvery scales.

rheumatoid arthritis a long-term condition that causes pain, swelling and stiffness in the joints.

T-cell a type of white blood cell (lymphocyte) that helps the immune system work properly to fight infection.

transcobalamin deficiency a disorder affecting the transport of vitamin B12.

tuberculosis (TB) a bacterial infection spread through inhaling tiny droplets from the coughs or sneezes of an infected person.

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.

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

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