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Common Variable Immunodeficiency (CVID)

What is CVID?

The main finding in CVID is of low levels of antibodies (also known as immunoglobulins) in the blood. The body uses antibodies to fight off infections, and in particular bacterial infections. CVID can affect both males and females and can begin at any age.

How is CVID diagnosed?

There are a number of causes of low immunoglobulins, and before CVID can be diagnosed these need to be excluded. The exact conditions which need to be checked for will depend on your symptoms, sex and age. Typical tests include:

- measuring the levels of immunoglobulins (there are 3 main classes called IgM, IgA and IgG)
- checking whether there are specific immunoglobulins present which can fight infection (we use tetanus, pneumococcus and haemophilus influenzae type b as indicators)
- if the specific immunoglobulins are low then the response of the immune system to vaccination is assessed
- counting the numbers of different immune cells in the blood

Why does it happen?

A type of immune cell known as B cells are responsible for the production of antibodies. In the immune system there is normally constant communication between B cells and other immune cells. The point of this communication is to ensure that B cells make antibodies against the correct targets. B cells help your immune system by making antibodies to fight against infections, and in particular bacterial infections. There are control mechanisms to stop B cells making antibodies against yourself, as this would cause autoimmune disease.
Why some people develop CVID is the subject of ongoing research and the causes are not fully understood. However, it appears that there is a problem in the communication between B cells and other immune cells. The effect of this is that the B cells stop producing good levels of antibodies against infections. Some, but not all patients with CVID also produce inappropriate antibodies and can develop autoimmune disease.

What might my symptoms be?

The low levels of antibodies result in increased susceptibility to infections. The usual pattern is an increase in the number and severity of bacterial infections of the lungs and sinuses. The symptoms result from the infections and may include cough, sputum production, fever, tiredness and muscle pains. Recurrent chest infections can cause damage to the lungs and can result in a condition known as bronchiectasis. In bronchiectasis sputum pools in the damaged areas and causes persistent coughing and also increases the chance of future infections. It is therefore important that chest infections are treated promptly to avoid the development or worsening of bronchiectasis.

What treatments can be used?

The main treatment for CVID is immunoglobulin replacement therapy. This replaces the low levels of immunoglobulins and can be administered in 2 ways. Intravenous immunoglobulin (IVIg) is given directly into the vein, usually every 3 weeks. Subcutaneous immunoglobulin (SCIg) is given just under the skin, for example on the abdomen, usually weekly. There is a newer method of administering immunoglobulin known as facilitated SCIg (fSCIg) which allows immunoglobulin to be given subcutaneously every 3 weeks. The choice between the different ways of infusing immunoglobulin will be discussed with you and the different methods suit different people. The dose and how often the infusions need to be given varies and is tailored to each individual's requirements.

Infections should be promptly treated with antibiotics. Often high dose antibiotics continued for at least 10 days are used. If the episodes of infection are frequent, you may be started on prophylactic antibiotics – this is a low dose of antibiotics taken regularly, even when you are well. The aim of prophylactic antibiotics is to prevent an infection from starting. The antibiotic chosen varies from person to person according to the individual circumstances. For example, allergies to antibiotics, the species of bacteria and the site(s) of infection will all influence the antibiotic choice.

Possible complications of CVID

Some patients with CVID develop autoimmune disease where the immune system attacks part of the body. There are a large number of different autoimmune diseases and the treatment will vary according to the condition. One example which is more common in CVID is low platelets. Platelets are needed for your blood to clot efficiently. We regularly check platelet numbers when you attend for clinic appointments.

CVID can also increase the risk of some cancers. This doesn't mean that if you have CVID you will develop cancer. What it does mean is that your chances are higher than that of the general population. Therefore, it is important that if you have concerning symptoms that they are reported to your doctor so that they can be investigated at an early stage. Such symptoms include a change in your bowel habit, passing blood, abdominal discomfort and unexplained weight loss. These symptoms would prompt us to perform tests so that any problems can be detected early and treated promptly.

Further information

CVID, as the name suggests, is variable. The above information is a summary of what affects CVID might have, but different patients have different symptoms. Please ask us in clinic any questions that you have about CVID and/or your treatment.

PID UK (Primary Immunodeficiency UK) and UKPIPS (UK Primary Immunodeficiency Patient Support) are patient groups for those affected by immunodeficiencies. Their websites are below (note that we have no editorial control for the websites and take no responsibility for their content):

www.piduk.org
www.ukpips.org.uk

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