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Hyper IgM

What is hyper IgM?

Antibodies (also known as immunoglobulins) circulate in your blood and are present in secretions such as saliva and lung secretions. They help the immune system fight infections. There are several classes of immunoglobulin; IgM, IgA, IgG and IgE.

In hyper IgM there is a lack of IgA, IgG and IgE. The levels of IgM though are either normal or raised (hyper IgM is not the best description of the disease as in some individuals the levels of IgM are normal). The lack of IgG in particular means that there is an increased susceptibility to infections.

Hyper IgM is an inherited condition and symptoms usually start from 6 months onwards. There are several types of hyper IgM and it can be inherited in two ways. The most common form is inherited as an X-linked condition. This means that it can only affect males, though females can be carriers. Rarer forms are inherited in an autosomal recessive manner. This means that it can affect both males and females, and that both males and females can be carriers. We can explain this further to you in clinic should you wish to know more.

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 should make antibodies against infections to help your immune system fight them off.

In hyper IgM this communication does not work. B cells cannot make the usual range of antibodies, though they are still able to produce IgM. Often, the levels of IgM are raised as this is the only class of antibody that can be produced.

During a bacterial infection IgM is the first line of defence, but after a few days other antibodies and in particular IgG are produced. Though it takes the immune system longer to start producing IgG, once produced it is more effective at killing bacteria than IgM. IgG also is important for the immune system's 'memory'. If on another date the same bacteria tries to cause a second infection, then this time IgG can be produced from the very beginning and prevent infection. The lack of IgG in hyper IgM means that there is an increased susceptibility to infections.

What might my symptoms be?

The low levels of antibodies results 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 results 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.

How is hyper IgM diagnosed?

The following tests are normally performed in investigation of hyper IgM:

- measuring the levels of immunoglobulins (there are 3 main classes called IgM, IgA and IgG)
- counting the numbers of different immune cells in the blood – these should be normal in hyper IgM
- looking for the presence of a molecules involved in the communication between B cells and other immune cells (one example is a molecule called CD40L). Sometimes genetic tests may be performed as these can help with the diagnosis.

What treatments can be used?

The main treatment 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 individuals 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

Further information

Please ask us in clinic any questions that you have about hyper IgM 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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