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Job's syndrome (Hyper IgE syndrome)

What is Job's syndrome?

Job's syndrome is a rare condition which affects both males and females with symptoms usually beginning in childhood. The most common features are eczema, increased susceptibility to infections and markedly raised levels of IgE (one of the classes of antibodies) on blood testing. Often the IgE levels are greater than 10,000 kU/l when the normal range is less than 81 kU/l.

Why does it happen?

Some cases of Job's syndrome have been found to be due to a genetic defect in a protein called STAT3. This protein is involved in passing signals from the outside of cells to the inside. Without STAT3, the cells are unable to respond properly to what is going on around them. Rarer causes of Job's syndrome are genetic defects affecting DOCK8, TYK2 and SPINK5. The other causes of Job's syndrome are not known, but are the subject of ongoing research.

What might my symptoms be?

Some symptoms are very common in Job's syndrome, whereas others are variable between different individuals.
The following are nearly always present: eczema, increased rates of infections especially lung infections, raised IgE levels.
The following are sometimes present: skeletal abnormalities such as increased risk of fractures, hyperextensible joints, dental abnormalities.

How is Job's syndrome diagnosed?

There is no single test for Job's syndrome, but the combination of the above features would lead to a diagnosis of Job's syndrome. Advances in genetics now allow for testing for some of the genetic disorders which can lead to Job's syndrome.

What treatments can be used?

Antibiotics are the first line of treatment. The choice of antibiotic and how often it is taken is tailored to the individual. 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.

Some patients with Job's syndrome, while having high levels of IgE (immunoglobulin type E), have low levels of some or all of the other immunoglobulins. These are types M, A and G, known as IgM, IgA and IgG. These immunoglobulins are important in helping the immune system fight off infections and in particular bacterial infections. If the levels of these immunoglobulins are low then you may be given 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. For those patients on immunoglobulin replacement, antibiotics are still used and remain a crucially important part of treatment.

Further information

Job's syndrome is variable between patients. The above information is a summary of what effects Job's syndrome might have, but different patients have different symptoms. Please ask us in clinic any questions that you have about your condition 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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