

# Immunology

## Your guide to understanding secondary immunodeficiency

### What is immunodeficiency?

The immune system helps to protect us from infection - when this system fails we call the condition an immunodeficiency. Immunodeficiency is classified as being primary when it is not the result of a disease or treatment that damages the immune system. Primary immunodeficiency is very rare and usually lifelong. Immunodeficiency is classified as being secondary when the immune system is damaged by a medical treatment or another illness i.e. it is secondary to something else. Secondary immunodeficiency can be a short, medium or long-term condition.

Secondary immunodeficiencies can occur from many causes including:

- Treatment for certain cancers (lymphoma, lymphocytic leukaemia) by radiation, drug therapy or bone marrow transplant for cancer.
- Treatment by bone marrow transplant for other non-cancer conditions such as metabolic disease.
- Treatment for autoimmune diseases.

Secondary immunodeficiency is a result of failure of components of the immune system that are mainly involved in protecting against infection. This may be a failure of production of immune cells or, although cell numbers are normal, they do not function well. When you are investigated to see whether you may have secondary immunodeficiency, we look at the numbers and types of immune cells in your blood and we measure the quantity and quality of the immune proteins (antibodies)

that these cells produce. Sometimes we will offer you vaccination to look at how well your immune system learns and remembers how to fight common infection. Antibodies are members of a family of proteins called Immunoglobulins (Ig) and are important in helping to fight infections, especially bacterial infections.

Secondary immunodeficiency means that you may be more likely to catch more infections than is normal. You have been referred to a Consultant Immunologist to investigate this.

### What treatment is available?

If our tests suggest that you do have an immunodeficiency, you may need support through antibiotics either regularly to prevent infection or to keep at home and use promptly at the first sign of infection. For some patients, however, this is not enough to keep them well. Patients who lack the right immunoglobulins to fight infections may be given replacement therapy via an infusion of immunoglobulins which 'tops up' their immune system. This is called immunoglobulin replacement therapy. Depending on the cause of the secondary immunodeficiency, you may need immunoglobulin replacement therapy for months or years or even for life. This will be discussed with you when you see the Immunologist in clinic and will be reviewed on a yearly basis. When a decision is made to review whether you are still likely to benefit from this treatment, you may have a planned treatment interruption or 'treatment holiday' whilst we do some blood tests and perhaps some vaccinations to see whether you still have an immunodeficiency.

## Immunoglobulin replacement therapy

This can be given into the vein, called intravenous immunoglobulin (IVIg), or into the subcutaneous fat of the skin usually in the abdomen or thigh called subcutaneous immunoglobulin (SCIg).

Immunoglobulin replacement therapy can be given in the day case unit. If it is given in hospital, it is most frequently given straight into your blood stream via an intravenous drip of immunoglobulin (IVIg). It takes a few hours and should not be painful. Your specialist centre can provide training so that you are able to carry out treatments in your own home, with someone to help you. Immunoglobulin can also be given subcutaneously (SCIg) (just under the skin). This makes it simpler to administer yourself at home. However, this process needs to be repeated more frequently than IVIg, as smaller volumes are administered each time. Your Immunology team will discuss with you the best way for you to receive your treatment, so that you can continue to live your life with as little disruption as possible.

This regime should keep most infections at bay and dramatically reduce the time spent off school or work due to sickness. Although the treatment cannot reverse damage already caused by severe recurrent infections, it will prevent more problems developing. Royal Papworth Hospital has a recognised home therapy training centre.

Side effects from either IVIg or SCIg are rare and are most likely to happen during the first few infusions.

## Other medicines which may be prescribed to you

Even on adequate treatment there may be 'breakthrough' infections. When these occur you must begin antibiotic therapy immediately. As soon as you begin to feel ill you must contact your GP to obtain a prescription for

antibiotics. The surgery staff should be made aware of your condition and ensure that your request is given priority.

## Safety of immunoglobulin

Immunoglobulin is prepared from human blood. There is potentially a risk of infection from blood-borne viruses such as Hepatitis and HIV (the AIDS virus). However, there have been **no** cases of HIV or Hepatitis B being transmitted in this way. All blood donations are screened for Hepatitis B & C, HIV, and new variant CJD and the purification stages reduce the possibility of infection to a minute level.

There are several preparations available, and your Consultant Immunologist will decide which one is best for you. Changing preparations is not advised under normal circumstances.

## Specialist care

Ideally patients with secondary immunodeficiencies should be looked after at a centre specialising in this condition. The Immunology Department at Royal Papworth Hospital is a specialist centre for diagnosis and management, and it is very important that you are seen regularly by its team of experts.

## Physiotherapy

Some patients with secondary immunodeficiencies may have suffered many chest infections, which may have damaged their lungs. If this is the case, you may be referred to a physiotherapist who will teach you exercises to help your breathing.

## Alternative therapies

No 'alternative' therapy can affect your ability to make effective antibodies. However, in theory, something which helps you to relax and feel good in yourself is unlikely to harm you. But please, consult the Immunology team before embarking on any new treatment. And remember, it is vital that you **always** receive regular immunoglobulin replacement therapy.

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Author ID: Immunology Nurse Specialist  
Department: Chest Medical Unit  
Printed: April 2019  
Review due: July 2020  
Version: 3  
Leaflet number: PI 77

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## Who do I need to tell?

Your GP will be kept informed of your progress and treatment. However, because it is rare, your doctor may not be fully familiar with your condition. More information and advice is always readily available from the team at Royal Papworth Hospital.

If you are referred for any surgery, you must inform the surgeon of your condition, and tell the Immunology team what is happening. This is because you are more susceptible to infections than an average patient, and your antibiotic cover and/or your immunoglobulin therapy may need to be increased to cope with surgery.

It is also a good idea to tell your dentist of your condition, and, if you have an accident, the Accident and Emergency Department.

## Immunisations

Regular replacement immunoglobulin therapy will keep you supplied with antibodies against most diseases. This replaces the need for most vaccinations and if you have been prescribed immunoglobulin, you are unlikely to benefit from immunisation. Although such vaccines contain an organism that has been treated to make it harmless to people with a normal immune system, they could adversely affect you.

The exception is the flu vaccine. As the flu virus changes every year, your immunoglobulin product may not provide adequate protection and you may derive some benefit from the vaccine. This is an inactivated vaccine. Generally you should not receive live vaccines such as MMR and yellow fever.

## Your lifestyle

- It is important that you look after yourself with a sensible diet and exercise. Swimming, cycling and walking will all improve your general state of health. If you were diagnosed before major problems occurred, then you can look forward to a normal lifestyle and to a normal life span.
- As chest infections are a particular problem, it is **vital** that you do not smoke, and that you ask **others** not to smoke around you and to allow you to live in a **smoke-free environment**.
- If you intend to have children, you must discuss this with your Immunologist, to obtain the best possible advice and treatment.
- You are more at risk of getting food poisoning and so **good** kitchen hygiene is **vital**.
- Pets should not present a health problem, as long as normal care is taken.
- Once you are on regular immunoglobulin replacement therapy, you should have fewer interruptions to your work or school due to illness; and home therapy makes life even easier.
- Young patients can go away to college as long as treatment at a local Immunology Centre is arranged.

## Travel

Holidays and business trips abroad can be undertaken, and even far-flung exotic destinations can be enjoyed, as long as advice is sought and followed.

## On-going support and further information:

For more information please ask your Specialist Immunology Nurse:

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