

Secondary Immunodeficiency

A patient's guide and
consent form

What is immunodeficiency?

The immune system helps to protect us from infection - when this system fails it may lead to illness including infection. When the immune system does not work correctly, we call the condition an immunodeficiency. 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

Most patients who have Immunoglobulin replacement therapy learn to treat themselves at home however for some patients, it is better to have the treatment in hospital. Your immunology team will discuss the treatment options with you.

If you want to learn to do your own treatments and minimize hospital visits, your immunology team will provide training so that you can carry out treatments in your own home, with someone to help you. Your treatment will be given subcutaneously (subcutaneous immunoglobulin, SCIG by injection just under the skin every week. This takes a few minutes.

If it is better for you to have your treatment in hospital, this will be given through a drip into your vein (intravenous immunoglobulin, IVIG) every 4 weeks in the ambulatory care unit. It will take approximately 5 hours.

Once you start regular immunoglobulin treatment, this should keep most infections at bay and dramatically reducing the time spent off work due to sickness. Although the treatment cannot reverse damage already caused by severe recurrent infections, it will prevent more problems developing. Side effects from either IVIg or SClg 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. Please also let us know. The immunology team operates a patient advice and support service:

Email: papworth.ldcicns@nhs.net;
Contact via switchboard: 01223 638000;
The service operates from 09:30-12:30,
Monday to Friday (excluding bank holidays).

Please see the patient information leaflet: Lung defence and respiratory immunology: patient advice and support service' for more information. This can be found on the public website.

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. Once established on a preparation, we try not to change unless there is a change in supply, and we need to change product to ensure that you continue to receive regular treatment.

Specialist care

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. Depending on your personal situation, we may work closely with your local hospital team and with your GP to make sure you have the best care.

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 and clear your lungs of congestion.

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. Please, consult the Immunology team before embarking on any new treatments. And remember, it is vital that you always receive regular immunoglobulin replacement therapy.

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 when you come to your clinic appointments and through the lung defence and respiratory immunology patient advice and support service.

Please affix patient label or complete details below.

Full name:

Hospital number:

NHS number:

DOB:

PIC 77: patient agreement to PI 77 - Immunoglobulin replacement therapy

Intended procedure/surgery

Statement of healthcare professional

(To be filled in by healthcare professional with appropriate knowledge of proposed procedure, as specified in consent policy).

I have explained the procedure to the patient. In particular I have explained:

The intended benefits:

- To replace immunoglobulin that the patient does not make.
- To prevent infections.

Significant, unavoidable or frequently occurring risks:

- Immunoglobulin is a human blood product, and there is therefore a risk of infection. This is eliminated as far as possible by donor screening for Hep B, C and HIV and the manufacturing process which inactivates infective agents. Immunoglobulin is sourced from the UK, EU and USA. However, the potential risk of acquiring infection cannot be completely eliminated.
- Adverse reactions can occur during the infusion, such as headaches, shivering, wheezing. These can be managed by stopping/slowing the infusion or giving appropriate medications.

Additional risks specific to you or your operation - please specify below:

.....

Any extra procedures, which may become necessary during the procedure:

Blood transfusion

Other procedure - please specify below:

.....

I have also discussed what the procedure is likely to involve, the benefits and risks of any available alternative treatments (including no treatment) and any particular concerns of this patient.

Healthcare professional

Signed:

Date:

Name (PRINT):

Job title:

Contact details

.....

.....

Has a ReSPECT form been considered and, if relevant, appended to this form? (See page 10 for further details on ReSPECT)

Yes

No

Statement of patient

Please read the patient information and this form carefully. If the treatment has been planned in advance, you should already have your own copy of which describes the benefits and risks of the proposed treatment. If not, you will be offered a copy now. If you have any further questions, do ask - we are here to help you. You have the right to change your mind at any time, including after you have signed this form.

Yes No

I agree to the procedure or course of treatment described on this form and have read this information leaflet on insert title (PI 77) and had the opportunity to ask questions.

I agree to the use of photography for the purpose of diagnosis and treatment and I agree to photographs being used for medical teaching and education.

- **I understand** what the procedure is and I know why it is being done, including the risks and benefits.

Please affix patient label or complete details below.

Full name:

Hospital number:

NHS number:

DOB:



Royal Papworth Hospital

NHS Foundation Trust

- **I understand** that any tissue removed as part of the procedure or treatment may be used for diagnosis, stored or disposed of as appropriate and in a manner regulated by appropriate, ethical, legal and professional standards.
- **I understand** that any procedure in addition to those described on this form will be carried out only if necessary to save my life or to prevent serious harm to my health.
- I have listed below any procedures **which I do not wish to be carried out** without further discussion:

.....

.....

.....

.....

I have been told in the past by Public Health that I am at increased risk of CJD (Creutzfeldt Jakob disease) or vCJD (variant Creutzfeldt Jakob disease).

Yes No

(Where patient indicates 'yes' health professional to refer to Trust CJD procedure DN092)

Statement of interpreter (where appropriate)

If an interpreter was present to support this consent, please state the name and number of the interpreter present:

Date:

Interpreter's number:.....

Name (PRINT):

If a telephone / video service has been used, please document the name of the interpreter and company below

.....

.....

.....

Patient

Patient signature:

Date:

Name (PRINT):

Confirmation of consent

(To be completed by a health professional when the patient is admitted for the procedure, if the patient has signed the form in advance).

On behalf of the team treating the patient, I have confirmed with the patient that they have no further questions and wish the procedure to go ahead.

Signed:

Date:

Name (PRINT):

Job title:

Important notes (tick if applicable).

- Patient has advance decision to refuse treatment
- Patient has withdrawn consent (ask patient to sign/date here)

Patient signature:

Date:

Name (PRINT):

Please use and attach Consent form C for a young person who is not Gillick competent.

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 immunoglobulin replacement therapy will keep you supplied with antibodies against most diseases. This replaces the need for vaccinations, and you should not receive any 'live' vaccines. Live vaccines contain an organism that has been treated to make it harmless to people with a normal immune system however they could adversely affect you. Some vaccines are recommended. These are non-live vaccines that may boost your immunity to help protect you from particular infections. The vaccines we recommend include the annual influenza vaccine, the RSV vaccine if you are in the appropriate age cohort (currently 75-80 years however this may start at a younger age in due course) and the SARS-CoV-2 vaccines. If you are unsure, please ask your immunology team.

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. A travel information leaflet is available to provide information relevant to keeping you well during your visit. Please ask your specialist immunology nurse for a copy of this.

On-going support and further information:

For more information, please ask your specialist immunology nurse:

Email: papworth.ldcicns@nhs.net;

Telephone: 01223 638000;

The service operates from 09:30-12:30, Monday to Friday (excluding bank holidays).

Patient organisation:

A comprehensive range of information resources for patients may be found through the national patient charity: Immunodeficiency UK

immunodeficiencyuk.org/resources/

The charity may be also contacted by email: hello@immunodeficiencyuk.org or by telephone: 0800 987 8986

Recommended summary plan for emergency care and treatment (ReSPECT)

What is ReSPECT?

ReSPECT stands for 'Recommended summary plan for emergency care and treatment'. It is a process that helps people to think about what treatment is suitable in an emergency, should they be unable to make decisions at the time.

Why is it important?

We know that, when people are very unwell, they are often unable to think clearly about what treatment they may or may not want because their brain and body are overwhelmed by the illness. It is also normal for people to feel anxious about what is happening when they are sick and in hospital, and this can also make it difficult to think clearly. This is why we think it is a good idea, where possible, for decisions about medical treatment to be made in advance – before there is an emergency situation or crisis.

How does it work?

The ReSPECT process is designed to help conversations between you and your healthcare professionals: they need to make sure you understand your health problems and which treatments may or may not benefit you. You need to make sure the healthcare professionals understand what matters most to you and whether there is anything you are particularly worried about or would want to avoid.

This conversation is used to complete a ReSPECT form that records a person's health problems, their preferences and which medical treatments may or may not be suggested. The original form should stay with the patient, though it is extremely helpful to have a record of the content of the form on their electronic patient record.

A ReSPECT form is NOT a legally binding document and can be changed or withdrawn at any point.

The ReSPECT form is often used to indicate treatments that someone may not want and/or treatments that their healthcare professionals consider would no longer be of benefit to them.

If people are getting worse from progressive conditions, it may be helpful to consider in advance about things such as whether they would wish to go back into hospital and, if in hospital, what sort of treatments might or might not be helpful for them.

This often includes a decision on whether or not they should have attempted cardiopulmonary resuscitation (CPR) if their heart was to stop.

Who is it for / is this relevant for me?

This process has increasing relevance for people who have complex health needs, people who may be nearing the end of their lives and those who are at risk of sudden deterioration or cardiac arrest.

However, many people come to Royal Papworth to have major procedures or surgery with the intention of curing a progressive disease or with the intention of substantially prolonging their life and, if that is you, you may wonder how a ReSPECT discussion applies to you and others like you. One of the key things to understand about the ReSPECT process is that it can be used simply to document a person's wishes and priorities, without setting any limitations on what treatment they should have.

This is important because all the procedures and operations we do here come with the risk of complications. In the unlikely event that things do not go as planned, it is really helpful to have some idea about a person's preferences and about their fears, worries and hopes.

Once again, the document is not legally binding, but it can help those looking after you to know what you might want if you weren't able to say for yourself.

The ReSPECT form is a separate document.

It is possible that your clinical team will start a conversation about the ReSPECT process with you. Equally you can ask any member of your medical team if you would like to start this conversation yourself.

Royal Papworth Hospital NHS Foundation Trust

A member of Cambridge University Health Partners



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Alternative versions of this leaflet

Large print copies and alternative language versions of this leaflet can be made available on request.

View a digital version of this leaflet by scanning the QR code.



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Scan the QR code or head to royalpapworth.nhs.uk/membership to find out more.



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