NHS
Royal Papworth Hospital
NHS Foundation Trust

Understanding primary immunodeficiency

A patient's guide and consent form

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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.

Primary immunodeficiencies are very rare diseases and some are so unusual that they only affect one or two people in the world. Any immunodeficiency means that you are likely to catch rather more infections than is normal. This is probably why you have been referred to a consultant immunologist.

Some immunodeficiencies are so severe they are diagnosed soon after birth. But others leave you at risk to just one or two types of infection, and it could be many years before you become sick. In addition, because these diseases are so rare, many doctors do not take them into consideration when diagnosing possible causes of recurrent, severe, or unusual infections. As a result, it is possible to suffer long term ill health and even permanent damage, before you are diagnosed with immunodeficiency.

With early diagnosis and proper care, most people can enjoy a reasonable quality of life.

What causes primary immunodeficiencies?

The cause is frequently found to be a faulty gene. Some immunodeficiencies are caused by a single faulty gene and others by a combination of genes. The fault may be inherited, be a new error in one of your genes or it may have been inherited from one or both of your parents. Most commonly, a faulty gene does not matter, unless the same fault is inherited from both parents.

This is most common when cousins have children together. Information about gene errors that cause primary immunodeficiencies is increasing rapidly due to new genetic research. Your hospital doctor will be happy to discuss how this new information could affect your diagnosis and treatment.

Gene therapy

Where a single faulty gene is found to cause the immunodeficiency, gene therapy, which means a normal gene is transferred to a patient to replace a faulty gene, is the best treatment of all. The new gene can correct the immune deficiency and cure the disease. Currently, this therapy is available for a few defined immunodeficiencies. Bone marrow transplantation is a more common option for patients where the immunodeficiency is very severe.

There is no link between Primary Antibody Deficiency and the Acquired Immunodeficiency Syndrome (AIDS). Patients with primary antibody deficiency diseases are not infectious and once treatment is started, they should start to feel better and suffer fewer infections.

There are a whole range of different primary immunodeficiencies. Antibody deficiencies are the largest group of primary immunodeficiencies. Antibodies are a group of proteins also known as immunoglobulins. This is the main group of proteins which help kill bacteria. There are three types of antibodies called IgG, IgA and IgM which work in different ways to protect us.

Antibody deficiency can range from a complete inability to make antibodies of any type, to more subtle illnesses, where one type of immunoglobulin is missing. Sometimes it may be that normal levels of antibodies are present, but they are not able to attack certain groups of bacteria.

What treatment is available?

Regular antibiotics are commonly used to prevent infections in patients with immunodeficiency. Depending on your individual situation, this may be a daily or three times a week oral antibiotic taken by mouth. Sometimes you may be offered an antibiotic that you breath in using a nebulizer. This treatment is used to target particular lung infections. For patients with antibody deficiency, the best treatment is to be given replacement therapy. This means that you are given antibodies prepared from other people who produce effective antibodies. This is called immunoglobulin replacement therapy. The immunoglobulins are prepared from donated blood plasma and given at weekly or monthly intervals. This is generally a long-term treatment.

Immunoglobulin 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 specialist centre 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 reduce 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.

The objective of immunoglobulin replacement therapy is to keep your immunoglobulin levels in the normal range for your age and protect you from infections.

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. Please also let us know. The immunology team operates a patient advice and support service:

Email: papworth.ldciccns@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.

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 primary 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 of primary and secondary immunodeficiency and it is very important that you are seen regularly by its team of experts.

Please affix patient label or complete details below.

Full name:

Hospital number:

NHS number:

DOB:

PIC 76: patient agreement to PI 76 - 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 11 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 76) 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.

Top copy to be filed in medical notes, carbon copy to be retained in booklet for patient.

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 Jacob 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.

Top copy to be filed in medical notes, carbon copy to be retained in booklet for patient.

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be retained in booklet for patient.

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

Many people with primary immunodeficiencies are not diagnosed until they 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. 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 defencce and respiratory immunology patient advice and support service.

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 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 life stylelifestyle 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, please 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 due to illness.

- Young patients may go away to college with pre-arranged treatment support at a local immunology centre.

Travel

Holidays and business trips abroad can be undertaken and even far flung exotic destinations can be enjoyed. 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.ldciccns@nhs.net

Telephone: contact via switchboard: 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 which has an excellent web platform:

immunodeficiencyuk.org/resources/

The charity may be also contacted by email:
Email us at hello@immunodeficiencyuk.org
or by telephone: Call us on 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
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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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As a member of Royal Papworth Hospital Foundation Trust, you could have your say on how the organisation is run, now and in the future. You will receive regular information and news from Royal Papworth and get invited to exclusive events. Membership allows you to vote for your representatives on the Council of Governors, talk to your Council of Governor representatives and stand as a governor.

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

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