Immunoglobulin therapy

A guide for young people and their families

NHS
Great Ormond Street Hospital for Children
NHS Foundation Trust

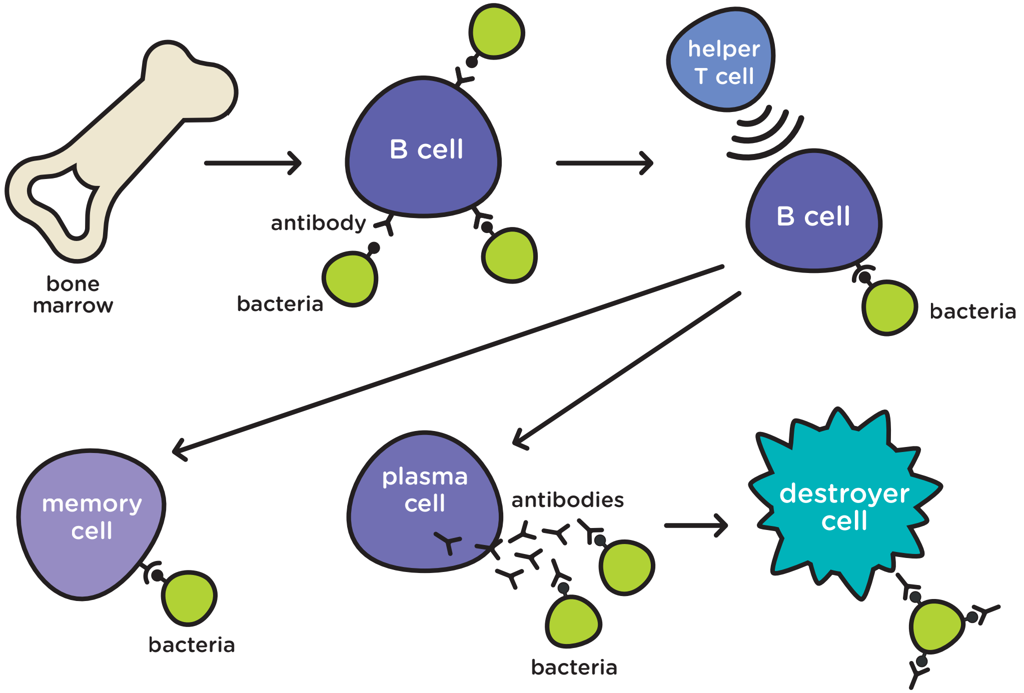
THE great north
CHILDREN'S HOSPITAL

IMMUNO DEFICIENCY UK

Supporting families affected by primary and secondary immunodeficiency

What is immunoglobulin?

Immunoglobulin (Ig for short, and sometimes called antibodies) is a part of the body's defence (immune) system. Your immune system helps to protect your body against infections that can be caused by different microbes, such as bacteria, viruses, parasites and fungi. Immunoglobulin treatment helps people who have immune systems that are weakened to fight off infections.



About this booklet

This booklet is written by Immunodeficiency UK, together with Great Ormond Street Hospital and the Great North Children's Hospital. It has information about immunoglobulin therapy (Ig therapy) for young people affected by primary immunodeficiency (PID) and secondary immunodeficiency (SID) and their families. There is a separate leaflet for younger children. The information should be used alongside guidance from your immunology team.

We would like to thank the young people and parents who helped to make this leaflet.

Immunoglobulin therapy – a guide for young people and their families
First edition August 2024

Published by Immunodeficiency UK (www.immunodeficiencyuk.org)

The immune system is a complex and clever function of our body's defence against infection. Each part has a special role to play and can interact with other parts to fight infection.

Immunoglobulin (which is also known as antibodies) is made by B cells. B cells come from the bone marrow, which is where all blood cells are made. T cells are a different type of blood cell. 'Helper' T cells have an important role in activating B cells when there is threat of an infection. When B cells are activated, some become plasma cells that produce more antibodies. Others become memory cells that recognise harmful substances that the body has seen before. This response helps to speed up the defences to deal with the threat of infection and prevents repeat infection.

Your doctor has decided you need immunoglobulin to keep you well.

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Your doctor and nurse will provide information on immunoglobulin treatment and the different options.

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You and your family will have a chance to ask questions and discuss what treatment option is best for you.

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Your treatment will be set up either in hospital, or at home after you/your family have had training.

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You will have regular follow-up with your doctor and nurse to see how well you are doing.

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You and your family will have ongoing support about your treatment.

Why is immunoglobulin needed?

You/your child will have had blood tests which show that the level of immunoglobulin is low. A low level of immunoglobulin is called hypogammaglobulinemia or antibody deficiency. There are many other tests that show how the immune system is working in more detail, and there are many reasons why the immunoglobulin level might be low. For example, it can be because you/your child have had treatment that affects the immune system (secondary antibody deficiency). Or it can be because there is some part of the immune system that is not working properly to make immunoglobulin (primary antibody deficiency). Sometimes a genetic/inherited variation in the cells is the cause of the deficiency; other times the exact cause is not known.

Whatever the cause, the treatment is the same; that is, to replace the immunoglobulin that is missing in your/your child's body. People with antibody deficiency are at a greater risk of infection than people with a fully functioning immune system. Research shows that with immunoglobulin treatment, however, fewer and less serious infections occur. Immunoglobulin treatment also helps to keep the body healthy for longer.

What is immunoglobulin replacement therapy?

Immunoglobulin treatment can only be given as an infusion – as a drip or injection, and not as a tablet. It is given either intravenously (into a vein, this is called IVIG) or subcutaneously (under the skin, this is called SCIG). The immunoglobulin is made from plasma that comes from a group of donors. It means that the infusion contains a mix of antibodies from many people, which will help to protect you/your child against different types of infections. Immunoglobulin is a blood-based treatment, so special care is taken to ensure that the medicine is safe. This includes careful screening of the donors and special ways of purifying the product.

Regular infusions are needed to keep good immunoglobulin levels in the body. Your immunology team will discuss with you the best way for this medicine to be given.

What is the difference between intravenous and subcutaneous immunoglobulin?

Benjy, aged 15, having facilitated SCIG at home.

Intravenous IG (IVIG)
IVIG is usually given in hospital. It involves having a cannula (a small plastic tube that goes into a vein) so that the immunoglobulin can be delivered straight into the bloodstream. The immunoglobulin is administered from a glass bottle through a thin plastic tube that goes via a pump to control the speed of the infusion. Each treatment usually takes between two and four hours. When you/your child are connected to the infusion, you/they will need to stay on the hospital ward and the nurses might make observations such as heart rate, temperature and breathing to make sure that everything is OK. It is a good idea to bring a book or a game (even schoolwork) to keep you or your child occupied while the infusion is running. Most young people who receive IVIG have their treatment at their local hospital, under a 'shared care' agreement with the immunology centre. Infusions are usually given every 3–4 weeks.

Subcutaneous IG (SCIG)
SCIG can be given in different ways. The first dose of SCIG needs to be given in hospital, but subsequent doses can be given at home when someone has been trained to give the medicine, such as a parent or a nurse. Young people also learn to do their own treatment. The infusions are given via a syringe that is connected to a thin plastic tube and two small needles that are put into each thigh, or each side of the abdomen (where the skin is a little bit fleshier to absorb the medicine). SCIG infusions take about 45–60 minutes if given via a small pump. If a smaller dose is pushed by hand, then the process takes a few minutes. There isn't much room under the skin, so it is normal that you/your child will see a swelling after the infusion and sometimes it is also a little bit red. The swelling is not usually painful and it will go away after a few hours to a day as the medicine is absorbed. The small space under the skin also means that the dose of SCIG will need to be smaller than with IVIG and given more often. SCIG is usually given every 1–2 weeks.

There is a different type of SCIG that is called facilitated-SCIG and this can be given as a bigger dose and less often. This SCIG infusion is 'facilitated' by a medicine that is given just before the immunoglobulin infusion and which makes the skin more stretchy. This allows a bigger infusion and therefore treatment can be given every 3–4 weeks. Talk to your immunology team to find out if facilitated-SCIG is an option for you or your child.

Will there be a choice of treatment?

Your immunology team will give you all the information you and your family need to help you decide which treatment option is best for you.

You might want to consider the following factors:

- If you and your family want flexibility around when and where you do the treatments, then SCIG infusions at home will give you more control.
- If it is difficult to put a cannula into your/your child's veins, then SCIG infusions will be easier than IVIG.
- If you/your child would prefer not to have infusions at home or you do not want the extra responsibility of doing the treatment, then IVIG in hospital will be a better option.
- If you/your child need long-term treatment with immunoglobulin (more than six months, or life-long) then SCIG treatment will involve fewer hospital visits and impact less on school and/or work.
- If you/your child have had side effects with IVIG, then SCIG infusions may be better tolerated.

The treatment centre will usually be able to offer IVIG or SCIG depending on what suits you and your family best. Many children start on IVIG to get their immunoglobulin levels up quickly. When they and their family feel ready, they then switch to SCIG. However, SCIG can also be started from the outset.

Whatever treatment option is chosen, you/your child will be guided, trained and supported by your immunology team every step of the way.

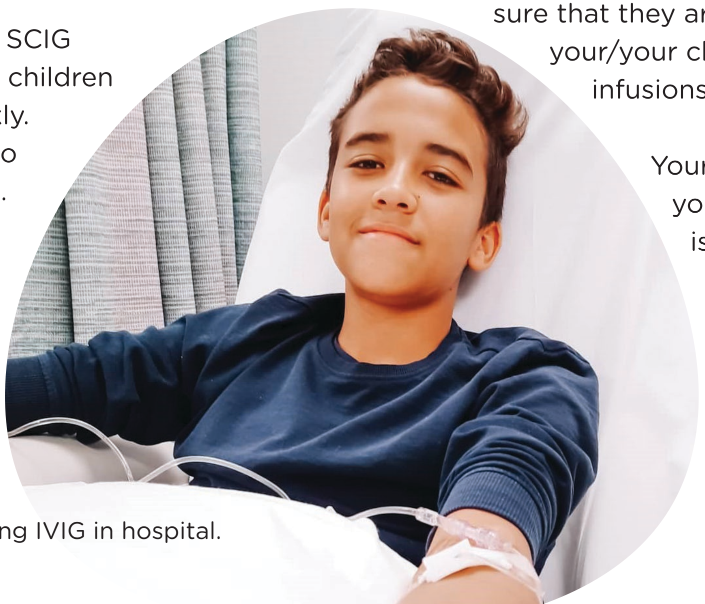
Are there any risks with the treatment?

Most people do not have any problems with their immunoglobulin treatment, but there are two sorts of risks that you should know about.

The first is that all medicines can have side effects (unwanted effects) as well as the good effects of protecting you from infections. Side effects could be things like a feeling of wanting to be sick, having a headache or a temperature. They could happen when you/your child are having the immunoglobulin treatment, but the person who is giving the treatment will be able to manage any side effect that happens. If you/your child have side effects with IVIG, then it is usually enough to slow the infusions down, and paracetamol can be given to lessen a headache or bring down a high temperature. It is more unusual to have side effects with SCIG because the infusions are absorbed slower when given under the skin than when given straight into the bloodstream. With SCIG infusions, you/your child can get a little bit of redness or itchy skin. There is medicine to help with this if that happens.

The second risk is catching an infection from your immunoglobulin treatment. This could possibly happen if any bacteria or viruses got into the medicine either when the medicine was made or when the infusion was administered. But the risk of this happening is incredibly small because all medicines are made and tested extremely carefully to make sure that they are clean and safe. The person who is doing your/your child's treatment will also know how to give the infusions in the correct way to avoid contamination.

Your team will monitor you/your child to check that your/their body is tolerating the treatment well. It is important to remember that the infusions are given to protect you/them from getting serious infections – the possible risks of treatment are small compared to the bigger, good effect that we see normally.



Jul, aged 16, having IVIG in hospital.

What kind of follow-up is needed with this treatment?

The exact follow-up plan varies between centres and will also depend on your/your child's condition. You are likely to see your immunology team at least two or three times a year. You will be asked to bring along the records of your infusions to your appointment. At the appointment, you/your child will be examined and asked health-related questions, such as:

Is the treatment working?

- Are infections still happening?
- Have any antibiotics been needed?
- Have days been taken off school/work or has there been a need to go into hospital?
- Is the right amount of immunoglobulin being given? (This is checked by doing a blood test.)

The doctor may check if your/your child's lungs are healthy by listening to the lungs with a stethoscope, and sometimes arranging a breathing test or a CT scan of the lungs.

Are there any problems?

- Have there been any side effects from the treatment?
- Are there any changes with the immune deficiency?
- Are there any new treatments or tests that should be considered?
- Is there an understanding of why you/your child is having immunoglobulin and are there any questions about the treatment or the immune deficiency condition?

Blood tests will be taken to check that there are no harmful effects on the liver or kidneys, and a sample might be frozen in case it needs testing for infection in the future. There might be a home visit if treatment is given at home.

At a follow-up visit, a large amount of information will be shared between you, your family and the immunology team. It can be difficult to remember everything that has been said. It is helpful to prepare for the appointment and to jot down any questions beforehand. Take notes or ask your team for some written information at your appointment.

Is it possible that immunoglobulin won't be needed anymore?

The tests you/your child had before starting immunoglobulin were designed to check whether you/they would need immunoglobulin for life: Sometimes immunoglobulin is recommended for people whose immune deficiency may be only temporary. This can happen in babies and small children or when the immune system has been damaged by medications. It is also possible that immunoglobulin was given for a condition that is no longer regarded as needing immunoglobulin. In these situations, blood tests will be done to check how well you/your child's immune system is working. If immunoglobulin therapy is stopped, your immunology team will monitor you/your child closely.

What immunoglobulin medicines are available?

There are about half a dozen companies that make immunoglobulin and their different IVIG and SCIG medicines vary slightly. The medicines all contain a similar mix of immunoglobulins, but they vary in the strength, the type of solution the immunoglobulins are mixed with and how they are stored. However, each company follows the same international standards on medicine safety.

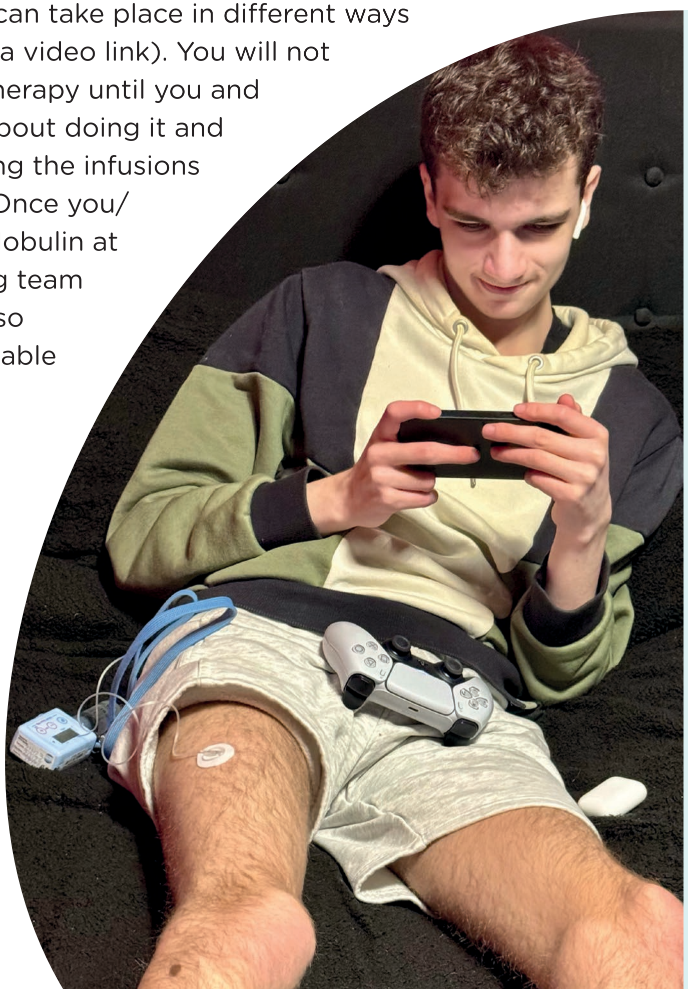
Kate, aged 14, having IVIG in hospital.

What if we want to go on holiday?

Immunoglobulin therapy should not affect your family's holidays. If you/your child are on IVIG, then a holiday of up to four weeks could be fitted into the infusion schedule. If weekly SCIG is given, then the infusion schedule could be adapted to allow a break for up to two weeks, or sometimes a single dose of IVIG can be given immediately before the holiday. Your immunology specialist nurse will help plan this.

How do I ensure that home therapy is being done safely?

Before starting treatment at home, you/your child will be fully trained. Your immunology specialist nurse will arrange this training with you and your family and it can take place in different ways (in hospital, at home and via video link). You will not be allowed to start home therapy until you and your family are confident about doing it and the person who will be doing the infusions is assessed as competent. Once you/your child are on immunoglobulin at home, the specialist nursing team will also do a review every so often, and they will be available for ongoing support and guidance if there are any questions or concerns.



Xander, aged 17, having SCIG therapy at home.

Who can I contact if I want to discuss my child's treatment?

Your immunology team will tell you who to contact if a problem arises at home; for example, if you think your child has an infection and you want advice about delaying the immunoglobulin treatment. Some immunology centres offer this service during office hours only. This means that you might want to plan your child's infusions for during the week until you have some confidence. You can also call your child's immunology centre if you have other concerns that might be to do with immune deficiency.

Your GP, NHS 111 and local emergency department will still be the best services to go to for all other problems, such as an injury.

Provision of immunoglobulin within the NHS

In the past, there have been times when the supply of immunoglobulin in the UK has fallen. This has happened if manufacturers have had to stop production for a few months or if manufacturers have got a better price for their product overseas.

Fortunately, these problems do not happen often, and they have never stopped patients in the UK getting the immunoglobulin they need. However, immunoglobulin is being used for more and more different illnesses, not just primary and secondary immunodeficiencies. As a result, contingencies to help safeguard supplies for people who need immunoglobulin have been put in place.

These include NHS systems to ensure that immunoglobulin healthcare needs are met and publication of clinical guidelines for immunoglobulin use.

Keeping infusion records (through hospital pharmacies or your own home therapy records) helps the national immunoglobulin database to be updated so that manufacturers can better manage supply and demand. Importantly these records also mean that any product issues with certain batches of immunoglobulin can be tracked directly to patients. This information is always anonymised.

Clinical guidelines for immunoglobulin use

Clinical guidelines ensure best practice in the use of immunoglobulin across all conditions requiring immunoglobulin. You can access and download the clinical guidelines for the different regions of the UK below:

England
Commissioning Criteria Policy for the use of therapeutic immunoglobulin (Ig) England, 2024 – **https://bit.ly/3LiSyNp**

Northern Ireland
In Northern Ireland, immunoglobulin is available as per the Department of Health guidelines, using an evidence-based prescribing process adopted from England. Northern Ireland enters individuals onto the demand management national database, which helps to provide a clearer picture across England, Scotland and Northern Ireland of the use of immunoglobulin in immunodeficiency conditions that are eligible for treatment with this therapy.

Scotland
The Commissioning Criteria Policy for the use of therapeutic immunoglobulin (2024) in Scotland is the same as in England – **https://bit.ly/3LiSyNp**

Wales
Wales has taken a different approach from the demand management programme in England. A professionally led All Wales Immunoglobulin Strategy Group has been set up and stocks are managed using a national Product Selection Guide. Responsible use of immunoglobulin products is very much a priority, and treatment decisions are clinician-led. Immunoglobulin products are managed via the Welsh Blood Service and local blood banks/pharmacies.

Immunoglobulin is often an effective way of treating immune deficiency.

Immunoglobulin replacement is always started and supervised by a specialist team.

Immunoglobulin is given directly into the body, either through the skin or into a vein.

Patients can have the treatment in hospital or, in many cases, can be trained to have the treatment at home.

Immunoglobulin therapy is safe but occasionally there are side effects that can be easily managed.

Careful review is provided by specialist immunology teams.

Top tips

Keep hydrated

It is good to drink a lot on the day when you have immunoglobulin treatment.

Get comfy

Wear comfortable and suitable clothes for having the infusions.

It’s good to talk

Explain your treatment to, and share your feelings with, people that are close to you – family, friends and your immunology team. They can listen and support you if things are sometimes feeling difficult, and they will want to know what you are going through.

Buddy up

Have an infusion buddy – someone who can accompany you to the hospital on the days you have infusions and who is able to help manage the information and what happens during the day. If a parent is doing your infusions at home, they might also want a buddy to help set things up and make them a cup of tea while the treatment is running.

Live well

Look after yourself, eat healthily and keep active – and the immunoglobulin will also help you to stay well.

Treat yourself

Make infusion day a treat day – do something nice while having the treatment or afterwards. Watch a film/play a game/have ice cream/go shopping/take a bike ride/play football/go to the cinema with a friend… What do you like doing?

Any questions?

Know how to contact your immunology team and get in touch with them if you have any questions or worries.

Glossary of terms

antibody – a type of protein (immunoglobulin) that is produced by certain types of white blood cells. Antibodies fight germs (mostly bacteria and viruses).

cannula – a small plastic tube that is inserted into a vein to give immunoglobulin or other treatments.

CT scan (also known as a CAT scan) – a specialised X-ray that gives pictures of the inside of the body.

immune deficiency – when the immune system's ability to fight infections doesn't work properly.

immune system – the parts of the body that protect against infection and disease.

immunoglobulins – proteins in the body that act as antibodies. They work to protect against and fight off infections. They are produced by specialist white blood cells and are present in blood serum and other body fluids.

immunologist – consultant specialising in the care of people with immune system problems.

infusion – delivery of treatment either into a vein (intravenous) or under the skin (subcutaneous).

intravenous – inside or into a vein; e.g. an immunoglobulin infusion may be given directly into a vein.

IVIG – intravenous immunoglobulin.

protein – one of the basic building blocks of life. Proteins make up the structure and determine the function of the cells that make up all the tissues of our bodies.

SCIG – subcutaneous immunoglobulin.

subcutaneous – meaning 'under the skin'. It also refers to anything relating to the loose tissue under the skin; e.g. an immunoglobulin infusion given straight into the tissue directly beneath the skin is said to be given subcutaneously.

Notes

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About Immunodeficiency UK

Immunodeficiency UK is a national organisation supporting individuals and families affected by primary and secondary immunodeficiency.

We are the UK national member of IPOPI, an association of national patient organisations dedicated to improving awareness, access to early diagnosis and optimal treatments for PID patients worldwide.

Our website has useful information on a range of conditions and topics, and explains the work we do to ensure the voice of patients with primary and secondary immunodeficiency is heard. If we can be of any help, please email us or call on the number above, where you can leave a message.

Support us by becoming a member of Immunodeficiency UK. It’s free and easy to do via our website. Members get monthly bulletins.

Immunodeficiency UK is reliant on voluntary donations. To make a donation, please go to **www.immunodeficiencyuk.org/donate**

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This booklet was made possible, in part, by a grant provided by the Jeffrey Modell Foundation WIN Program.