Subcutaneous treatment with gammaglobulin

Information for patients and their families
You or your child has been prescribed regular treatment with gammaglobulin, also known as immunoglobulin.

Here you will find a brief description of the immune system, immune deficiency disorders and Hizentra®, the gammaglobulin that your doctor has chosen to treat you or your child with.
The immune system is composed of several different elements that work together to protect your body from foreign infectious agents, such as bacteria or viruses. The non-specific (in-born) immune system which includes the skin and mucous membranes form the first line of defence against infectious agents. The specific immune system includes white blood cells and B-and T-lymphocytes.

**B-lymphocytes and antibodies**

B-lymphocytes make antibodies which take part in the body’s defences against infection, primarily against infections caused by bacteria. Each antibody binds to a specific infectious agent which it is specialised to recognise. Once this binding has taken place, the B-lymphocytes know which infectious agent is to be destroyed.

**T-lymphocytes**

T-lymphocytes are the most important in fighting viruses. They can also activate the B-lymphocytes and help them produce antibodies.

**There are several, different types of antibodies, including:**

**IgA**

IgA is present mainly in the body’s mucous membranes and prevents the infectious agent from penetrating further into the body.

**IgG**

IgG circulates in the blood and is also present in the tissues. IgG is divided into four subclasses: IgG1, IgG2, IgG3 and IgG4.

**IgM**

IgM circulates in the blood and is also present in the tissues. IgM is the first antibody to be formed when you have an infection.

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**Facts box**

Gammaglobulin = Immunoglobulin = antibodies
Immune deficiencies

Primary antibody deficiencies
Primary antibody deficiencies are the result of a defect in the immune system which affects the ability of the B-lymphocytes to produce sufficient amounts of antibodies, resulting in weakened defences against infections.

Secondary antibody deficiencies
Secondary antibody deficiencies occur as a result of a disease or medical treatment and is not caused by a defect in the immune system.

The most common antibody deficiencies

IgA deficiency
Deficiency of IgA is the most commonly occurring form of antibody deficiency.

IgG deficiency
Too little total IgG or too little of one or more of the IgG subclasses.

Variable immunodeficiency (CVID)
In variable immunodeficiency, CVID (Common Variable Immunodeficiency), there is normally too little IgG and IgA and, in some cases, too little IgM.

X-linked agammaglobulinemia (XLA)
X-linked agamma-globulinemia (XLA), only affects boys and is the result of a defect in the X chromosome. The defect gives a severely diminished ability to produce all types of antibodies.
Hizentra® is concentrated IgG and is extracted from blood plasma from a large number of healthy blood donors. The donors undergo a very thorough medical check-up and are tested for any infectious agents such as HIV and hepatitis viruses. During production, the plasma also undergoes various cleansing stages which inactivate any infectious agents.

Hizentra® is available in the following packs

![Hizentra® packs](image)

5 ml 10 ml 20 ml 50 ml

Hizentra® can be stored at room temperature for its entire shelf life. Hizentra® is also available in 10 x 10 ml packs.
Hizentra® replaces missing antibodies

The treatment of antibody deficiency involves regular administration of Hizentra®, normally once a week, to replace the antibodies that the B-lymphocytes in the immune system cannot produce. The purpose of the treatment is to reduce the number of infections.

Hizentra®, which is a solution for injection, is administered into the fat under the skin (subcutaneous) on the stomach, thigh and/or buttocks. This subcutaneous treatment is simple and safe, and generally you will administer it at home by yourself after receiving instruction and training from a nurse and doctor at your Specialist Department.

Gammaglobulin is injected here
How to administer Hizentra®

Instructions step-by-step
The Specialist Departments in Sweden may have different routines for the treatment, as well as different types of infusion aids i.e. pumps, draw-up cannulas, syringes, infusion needles, etc. This instruction leaflet illustrates a few types of infusion aids. If you have been prescribed other aids, follow the instructions you have been given by your department.

For children who need an anaesthetic cream or plaster, follow the instructions you have been given by your department.

1. Wash your hands with soap and water.

2. Collect together the right concentration of Hizentra® and the infusion aid you have been prescribed.
3. Check the expiry date on the vials. Check that the vials are intact and that the liquid is clear. Remove the cap seal, leaving the rubber membrane in place on the vial.

4. Prepare the infusion. Remove the protective cover from the draw-up cannula and place the vial on a flat surface. Insert the tip of the draw-up cannula through the vial's rubber membrane and open the cap on the draw-up cannula.

5. Attach the needle to the draw-up cannula.

6. Turn the vial and the draw-up cannula upside down. Make sure that the tip of the draw-up cannula is always covered by liquid. If you get bubbles in the vial, wait a few seconds and they will disappear. Fill the syringe by carefully pulling the plunger down. When the syringe is full, remove it from the draw-up cannula.
7. Remove the air from the syringe by pushing the plunger lightly until all the air has been expelled.

8. Attach the infusion needle tubing to the syringe. Press carefully until the gammaglobulin fills the tubing.

9. Clean the skin and stick the infusion needle into the fat under the skin on the stomach, thigh and/or buttock as you have been instructed.

10. Fix the needle in place using tape.
11a. It is very important that the tip of the needle does not enter a blood vessel. If this happens, you could experience severe side effects. **Check that the tip of the needle is not in a blood vessel by pulling back the plunger in the syringe.** If blood enters the tubing at the injection site, you must start again with a new infusion needle and new injection site.

11b. Remove the tubing from the needle for 15 seconds to check that no blood appears. Note that you must look for blood in the tubing close to the needle, not close to the syringe. If no blood appears in the tubing, you can continue.

12. Attach the syringe to the pump and start the treatment. When the infusion is finished, remove the infusion needle and apply a gauze pad or plaster. All materials used must be discarded in a safe manner according to the instructions you have been given by your nurse.
Side effects, warnings and precautions

**Side effects**
Serious side effects are uncommon, but local reactions in the skin at the injection site do commonly occur at the start of the treatment. Most common are swelling, tenderness, redness and itching. These local reactions are not harmful and diminish with time.

Fever, chills, headache, tiredness, migraine and allergic reactions also occur in individual cases. If you experience an allergic reaction during the treatment or immediately afterwards, you must contact the healthcare services as instructed by your nurse.

**Warnings and precautions**
Hizentra® must be administered subcutaneously into the fat under the skin and must never be injected into a blood vessel as this can result in effects on the heart and blood vessels such as causing a drop in blood pressure.

Clinical experience with gammaglobulins has not revealed any harmful effects on the unborn or newborn child, but talk to your doctor if you use Hizentra® when you are pregnant or breastfeeding.

For further information, see the package leaflet or go to fass.se.
Notes
You can find more information about the immune system and immune deficiencies here

PIO, Patient organisation for primary immunodeficiency
PIO is an organisation for people with primary immunodeficiency, their families, healthcare personnel and other interested parties.
www.pio.nu

The National Board of Health and Welfare website for rare diseases
www.socialstyrelsen.se/ovanligadiagnoser

International Patient Organisation for Primary Immunodeficiencies (IPOPI)
The international umbrella organisation for national organisations from around the world.
www.ipopi.org

Jeffrey Modell Foundation
A foundation providing information about primary immunodeficiency.
www.jmfworld.com

This leaflet has been professionally reviewed by:
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Hizentra® 200 mg/ml solution for subcutaneous injection. Human normal immunoglobulin (SCIg = subcutaneous immunoglobulin). Hizentra® is a medicinal product that is used to increase low levels of immunoglobulin in your blood to normal levels (replacement treatment). Do NOT inject Hizentra®: if you are allergic to human immunoglobulins, polysorbate 80 or L-proline; if you have hyperprolinaemia (a genetic disorder with high concentrations of the amino acid proline in the blood); into a blood vessel. Talk with your doctor or healthcare personnel before using Hizentra®. Read this package information leaflet carefully. CSL Behring AB, Tel. +46 8 544 966 70. This information is based on the package information leaflet 02/2016.

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