Hizentra®

Human normal immunoglobulin (SCIg) 20% solution

Subcutaneous immunoglobulin treatment for immunodeficiency

Information for patients and their relatives

Biotherapies for Life™ CSL Behring

You or your child has been prescribed
regular immunoglobulin treatment.

Here you will find a brief description of
the immune system,
immune deficiency disorders and Hizentra,
the immunoglobulin
that your doctor has chosen to treat
you or your child with.

The immune system

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 (inborn) immune system, which consists, for example, of the skin and mucous membranes, forms the first line of defence against infectious agents. The specific immune system consists, for example, of white blood cells and includes B- and T-lymphocytes.

B-lymphocytes and antibodies

B-lymphocytes form antibodies that assist with the body's immune defences, primarily against infections caused by bacteria. Each antibody binds to the specific infectious agent that it specialises in recognising. When this binding has taken place, the B-lymphocytes know which infectious agent is to be destroyed.

T-lymphocytes

T-lymphocytes are 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 – present mainly in the body's mucous membranes, this prevents the infectious agents from penetrating further into the body.

IgG – this circulates in the blood and is also present in the tissues. IgG is divided into four sub-classes: IgG1, IgG2, IgG3 and IgG4.

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

Fact box

Immunoglobulin = Gammaglobulin = Antibodies

Antibody deficiencies

Primary antibody deficiencies

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

The most common antibody deficiencies

IgA or IgG deficiency: These conditions occur when you have too little total IgA or IgG. With IgG deficiency, you may have too little of one or more of the IgG sub-classes.

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 agammaglobulinaemia (XLA): X-linked agammaglobulinaemia (XLA) only affects boys and is the result of a genetic defect in the X chromosome. The defect leads to a severely diminished ability to produce **all types of** antibodies.

Secondary antibody deficiencies

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

Hizentra®

Hizentra is concentrated IgG and is extracted from blood plasma from a large number of healthy plasma donors. The donors go through a very detailed medical check-up and are tested for any infectious agents such as HIV and hepatitis viruses. Also, during production, the plasma undergoes various cleansing stages that inactivate any infectious agents.

Hizentra is available in the following packs:

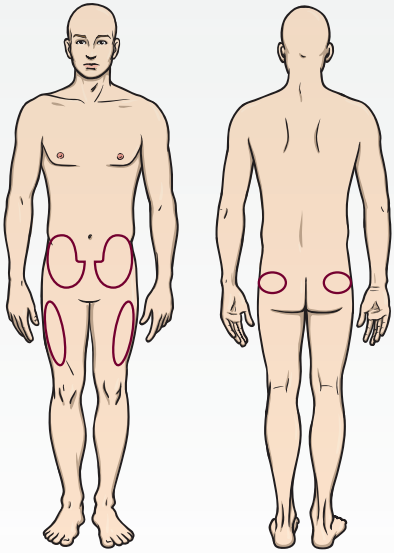
Hizentra can be stored at room temperature (up to 25°C) for its entire shelf life. Hizentra is also available in 10 × 10 ml packs.

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 (subcutaneously) on the stomach, thighs 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.

Immunoglobulin 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.

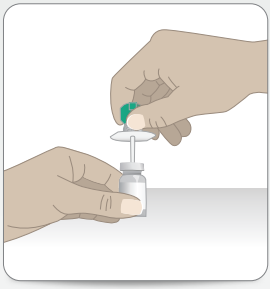
For children who need an anaesthetic cream or plaster, follow the instructions you have been given by your department.

1. Wash your hands with soap and water.

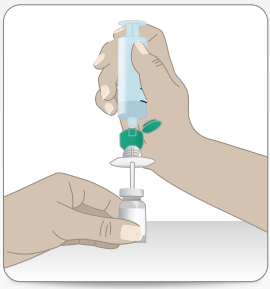
2. Collect together the right concentration of Hizentra and the infusion aid you have been prescribed.

3. Check that the expiry date on the vial has not passed, that the vials are intact and that the solution is clear. Remove the cap seal, leaving the rubber membrane in place on the vial.

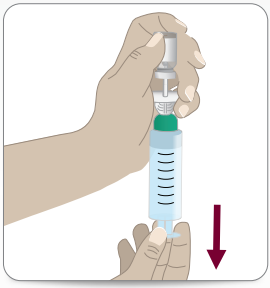
4. Prepare the infusion. Remove the protective cover from the draw-up cannula and place the vial on a flat surface. Insert the tip of the cannula through the vial's rubber membrane and open the cap.



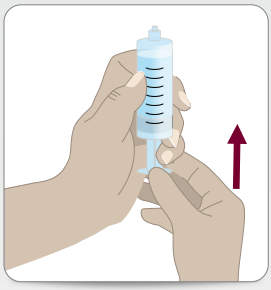
5. Attach the needle to the draw-up cannula.



6. Turn the vial and the draw-up cannula upside down. Make sure that the tip of the cannula is always covered by the solution. 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 the cannula.

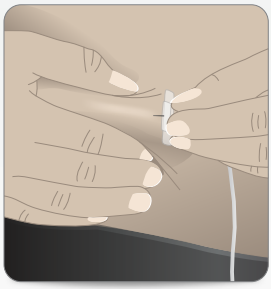


7. Remove the air from the syringe by pushing the plunger lightly until all the air has been expelled. Small air bubbles are not dangerous.

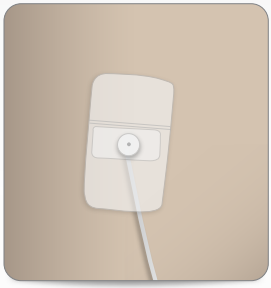


8. Attach the infusion needle tubing to the syringe. Press carefully until the immunoglobulin fills the tubing.

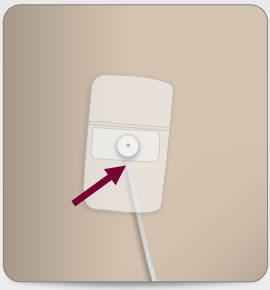
9. Stick the needle(s) into clean skin on the stomach, thigh and/or buttock as you have been instructed. There must be at least 5 cm between injection sites. Avoid inserting the needles into areas that are tender, bruised, red or hard. Avoid injecting areas that have scars or where the skin is broken.



10. Fix the needle(s) in place using tape.

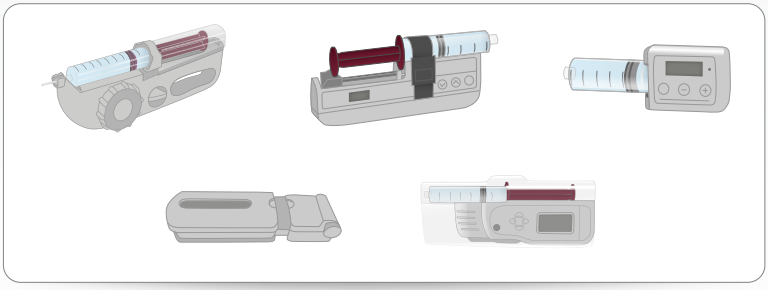


11a. It is very important that the tip of the needle does not enter a blood vessel. If this happens, you could experience severe side effects. **Check that the tip of the needle is not in a blood vessel by pulling back the plunger in the syringe.** If blood enters the tubing at the injection site, you must start again with a new infusion needle and new injection site. If an infusion set is used that has multiple needles, the tubing that is being used must be closed off using a clamp.

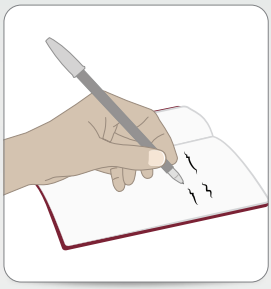


11b. Also remove the tubing from the needle for 15 seconds to check that no blood appears. Note that you must look for blood in the tubing close to the needle(s), not up at the syringe. If no blood appears in the tubing, you can continue.

12. Attach the syringe to the pump that you were prescribed and start the treatment.



13. When the infusion is finished, remove the needle(s) 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. Hizentra does not contain preservatives, so it is important never to reuse any solution that remains in the vials. Do not forget to document the batch number of the vials and the date and time of the infusion.



It is a good idea to refresh your memory from time to time by reading these instructions again.

Administration using manual pressure – so-called ‘rapid push’

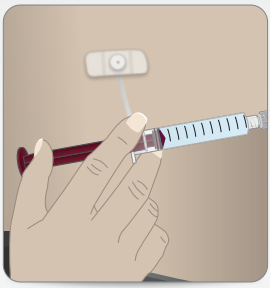
By using manual pressure – the so-called ‘rapid push method’ – you can control the infusion speed yourself rather than by using a pump.

You must perform your infusion in the manner described on page 7 and follow the instructions in the infusion guide, steps 1–11 and step 13. However, you can skip step 12 and therefore start your infusion straight after step 11.

Perform the infusion yourself as instructed by your department. They will also help you to choose a suitable needle for you, as well as telling you how many infusion sites to choose and how often to perform your treatment.

It is important to use only one infusion site per syringe. If you need to use another Hizentra syringe, use a new, sterile injection needle and change the infusion site.

Feel free to talk to your nurse if you would like more information about rapid push.



Side effects, warnings and precautions

Side effects

Serious side effects are uncommon, but local reactions in the skin at the injection site are very common 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. 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 a drop in blood pressure.

Clinical experience with immunoglobulins 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 Patient Information Leaflet in the package or go to fass.se.

Notes

You can find more information about the immune system and immune deficiencies here

PIO, Patient organisation for primary immunodeficiencies
PIO is an Swedish organisation for people with primary immunodeficiency and their families. www.pio.nu

Website of the National Board of Health and Welfare 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

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