

Your guide to self-infusing Aspaveli® ▼ (pegcetacoplan)

If you have any questions about your infusion or need additional training on how to use Aspaveli, please ask your doctor or nurse, or homecare provider

▼ This medicine is subject to additional monitoring.
This will allow quick identification of new safety information.
You can help by reporting any side effects you may get.
You can report side effects by contacting your doctor, nurse or pharmacist or you can report side effects directly via the Yellow Card Scheme at www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store.

This guide is intended for patients who have been prescribed Aspaveli, and is initiated and funded by Sobi™. Please refer to the Aspaveli Patient Information Leaflet (PIL) provided by your doctor or nurse for further information. The PIL is also available at www.medicines.org.uk/emc.

The information provided in this guide is general educational information and does not take the place of professional medical advice. Always follow your doctor or nurse's instructions and talk with them about any questions or problems you have regarding your health and treatment.

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Your guide to administering Aspaveli

This booklet has been given to you because you, or someone in your care, has been prescribed Aspaveli (pegcetacoplan) as a treatment for paroxysmal nocturnal haemoglobinuria (PNH).

This guide provides information about the steps to self-infusing Aspaveli.

Please read the Aspaveli Patient Information Leaflet before self-infusing Aspaveli.

Your first Aspaveli infusion/s will be administered in the hospital, where you and/or your caregiver will receive training from your doctor or nurse on how to set up for your infusion, and how to use the infusion pump.

You will also be given additional support and training at home, and will then be able to self-administer Aspaveli at home or at a location that is suitable and convenient for you.

In addition to this guide to self-infusing Aspaveli, a self-administration video is also available to support you.

Please type the below web address into your internet browser or scan the QR code to view the Aspaveli self-administration video.

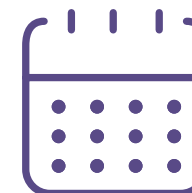
www.aspaveli-instructions.com/uk



Aspaveli is a treatment you administer at home or wherever is most convenient for you via subcutaneous (under the skin) infusion



The first time you are given Aspaveli, you will receive training on how to infuse it yourself, and ongoing training and support will also be available



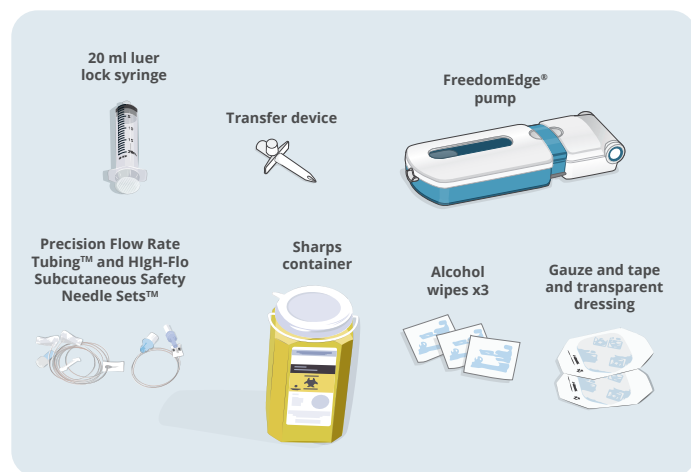
Aspaveli infusions should be done at regular intervals twice a week, as instructed by your doctor or nurse. In some circumstances, your doctor or nurse might change the frequency of your dose.



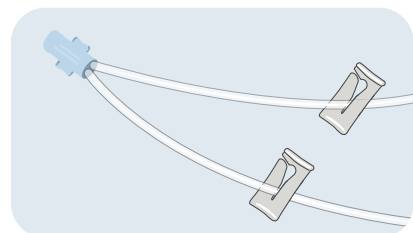
The typical infusion time is approximately 30 minutes when using two sites, or approximately 60 minutes if using one site for your infusion

Step 1: Prepare your infusion

- Wash your hands thoroughly with soap and water and dry them.
- Find a well-lit, flat work surface area (like a table).
- Thoroughly clean your flat work surface using an alcohol wipe.
- Remove a single-vial carton from the refrigerator. Keep the vial in the carton until it is ready for use to protect it from light and allow it to warm to room temperature (between 15°C and 25°C) for 30 minutes. DO NOT try to speed up the warming process.
- Gather all of the supplies that you need for your infusion (see image below), including the infusion pump, tubing set, needle set, and syringe and place these onto the cleaned work surface.



- The infusion should be completed within a maximum of 2 hours after withdrawing Aspaveli from the vial, or a new syringe should be prepared, using a new vial of Aspaveli. If a new syringe needs to be prepared because it has been more than 2 hours, contact your doctor or nurse, or homecare provider, to let them know.



- When you remove the needle set from its packaging, slide the plastic clamps across the tubing halfway down each side of the tubing.

Step 2: Check the vial and liquid

- Wash your hands thoroughly again with soap and water and dry them.
- After 30 minutes, remove the Aspaveli vial from the carton.
- Carefully inspect the liquid.
- Aspaveli should be a clear, colourless to slightly yellowish liquid – please check for any bits in the liquid or colour changes.



Do not use the vial if:

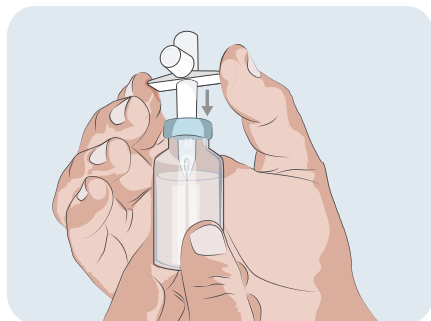
- The liquid looks cloudy, contains any bits, or is dark yellow
- The protective purple lid is missing or damaged
- The expiration date on the label has passed

Ensure that you prepare and carry out your infusion in an area away from pets and children.

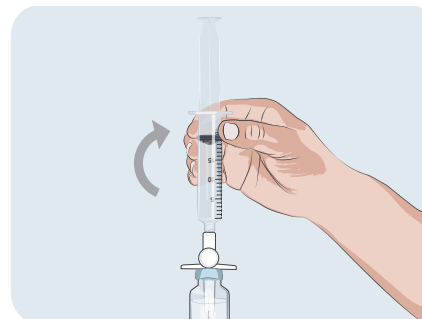
Step 3: Prepare and fill syringe



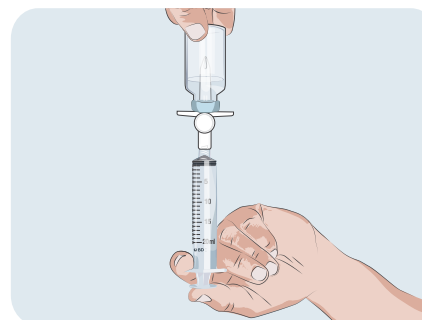
- Take off the purple lid from the vial to expose the grey rubber stopper.
- Use a new alcohol wipe to clean the top of the vial and let it dry for about 30 seconds.



- Place the vial onto your flat work surface area.
- Open the blister back and remove the vial transfer device.
- Remove the cover from the spike end of the transfer device, taking care not to touch the spike.
- Insert the spike straight down into the rubber stopper of the vial. Make sure the spike has penetrated the rubber stopper.



- Fill the syringe with 20 mL of air by pulling down on the plunger until the stopper reaches the 20 mL marking.
- Take this air-filled syringe and attach it to the top of the vial transfer device by pressing down firmly and twisting clockwise. Make sure it is securely attached.

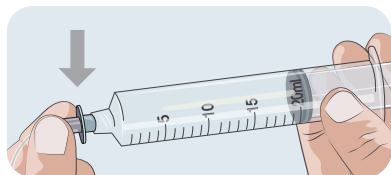


- Turn the vial upside down and slowly inject all of the air into the vial.
- Slowly pull the plunger to fill the syringe with all the Aspavali liquid from the vial.
- Ensure all the liquid has been transferred from the vial into the syringe.

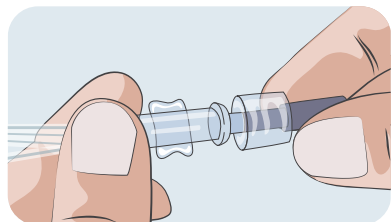


- Place the vial back onto your clean, flat work surface and remove the syringe by turning counterclockwise.
- Hold the syringe vertically upwards and push out any air bubbles from the syringe, taking care not to push out any of the liquid.

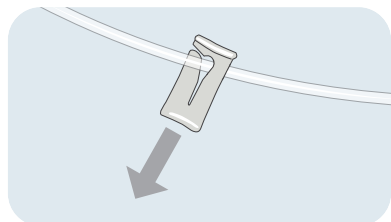
Step 4: Prepare the infusion pump and tubing



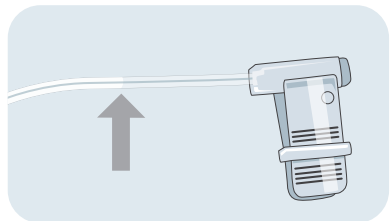
- Remove the cap from the luer disc end of the Precision Flow Rate Tubing™ set and connect the tubing to the syringe without touching the ends.



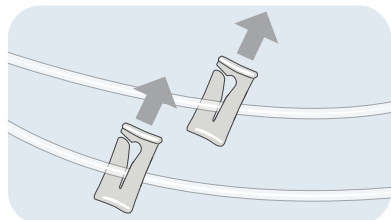
- Remove caps from the end of the High-Flo Needle™ set and the other end of the Precision Flow Rate Tubing™ set.
- Connect the sets together, taking care not to touch the ends.



- Unclamp one of the needle lines.

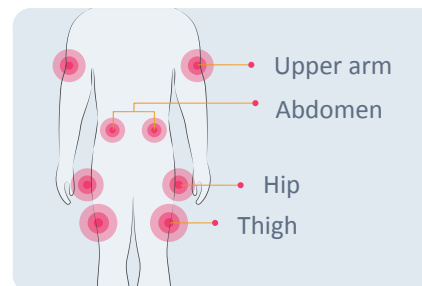


- Slowly and gently push the plunger of the syringe with steady pressure to fill the infusion tubing, it's important to go as slowly as you can. Doing this in front of a dark background can help you to see the fluid moving along the tubing. Stop when the liquid is about 2cm away from the needle as shown to you by your nursing team.

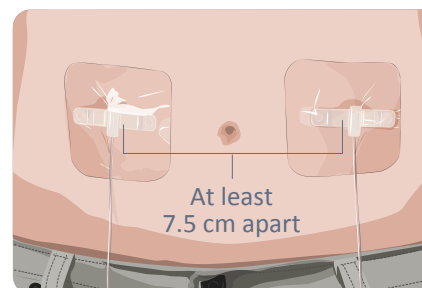


- Reclamp the tubing again halfway down the needle line.
- Repeat the above process for the second needle line.
- Once complete, make sure you have reclamped both needle lines.

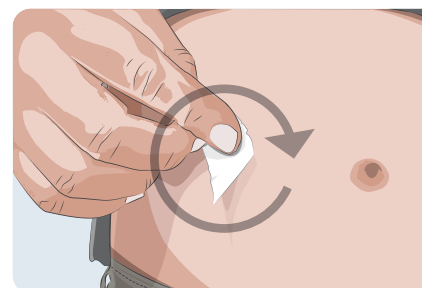
Step 5: Prepare the infusion site(s)



- Wash your hands again thoroughly with soap and water and dry them.
- Choose an area on your abdomen, thighs, hips, or upper arm for the infusion.
- Use a different site from the last time you infused Aspaveli and avoid any tender, bruised, red, or hard skin.



- When using two infusion sites, they should be at least 7.5 cm apart. If infusing in the abdomen, ensure that you pick a site at least 5 cm from your belly button.

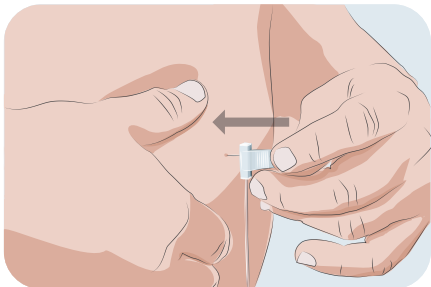


- Clean the skin at each infusion site with a new alcohol wipe, working outward in a circular motion.
- Let your skin dry for about 30 seconds.

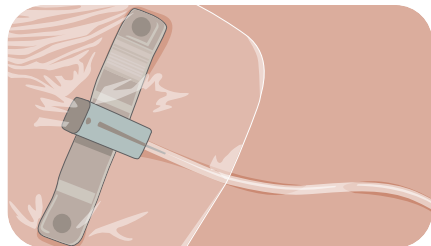
DO NOT INFUSE into the following areas:

- Tender, bruised, red or hard skin
- Tattoos, scars or stretch marks

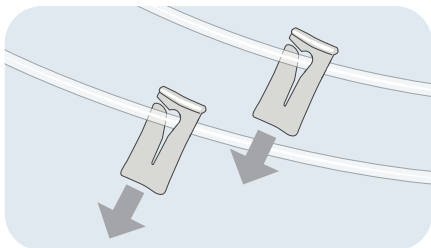
Step 6: Insert and secure the infusion needle(s)



- Without touching the needle, carefully remove the shield from the needle for the first infusion site.
- Pinch your skin between your thumb and forefinger around the infusion site (where you intend to place the needle).
- Insert the needle into the skin and fatty layer of tissue at a 90-degree angle, as shown in the image.



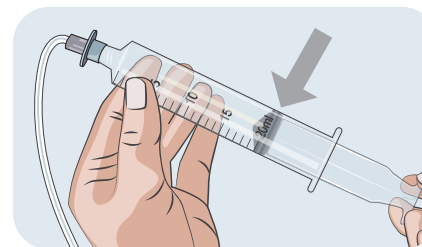
- Peel the printed side from the dressing to expose the sticky bit. Secure the needle with the dressing placed over the infusion site.
- Repeat this process for your second infusion site if you are using two sites.



- Unclamp both needle lines.

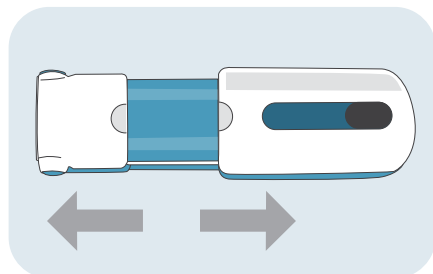
Step 7: Check for blood return (some blood coming into the needle and tubing)

At this stage, you may be trained by your nurse team to check for blood return. If so, please follow the instructions below. If not, please move to Step 8.

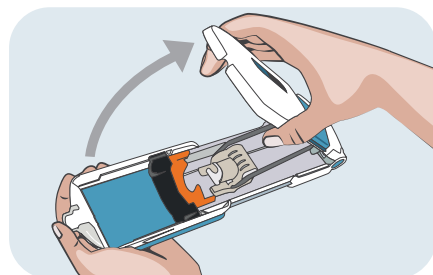


- Check for blood return by gently and slowly pulling back slightly on the syringe plunger.
- Watch to make sure no red or pink appears in the tubing near the needle(s).
- If blood return does occur, clamp the flow to that site and call your doctor or nurse to determine if the dose can be infused using the remaining site. If so, continue.
- If the dose cannot be infused using the remaining site, remove all needles and start again from step 4.

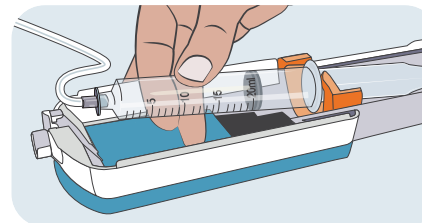
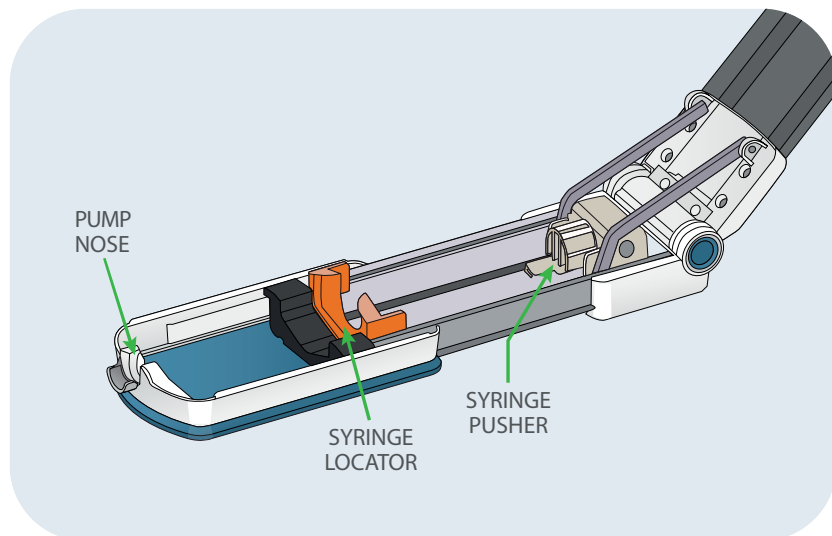
Step 8: Load the syringe and start the infusion



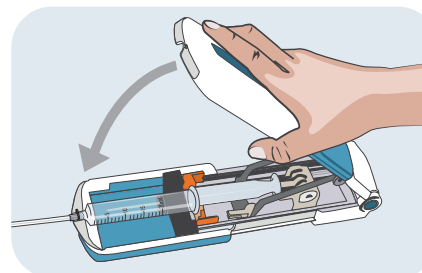
- To open the pump, pull firmly to fully extend it until you hear a click.



- Open the pump by lifting the top cover.
- The pump can be stiff to open. You may need to lift the top cover firmly to successfully open it.



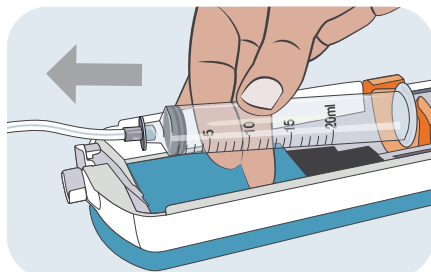
- With the lines and markings facing up, push the syringe against the orange syringe locator, plunger end first. If needed, pull back on the syringe locator to help.
- Seat the syringe flange (the widest part) into the syringe locator and seat the luer disc inside the pump nose so that the syringe is firmly attached inside the pump.
- You should not need to use lots of force to load or remove the syringe. You can test it fits by gently tugging on the syringe. It will stay in place if properly attached.



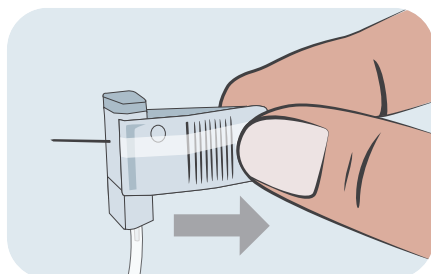
- Start the infusion by closing the pump. The infusion will begin immediately.
- To pause the infusion, open the lid to the pump. Restart the infusion by closing the lid.
- During the infusion, through the window of the pump, check to see that the syringe plunger is moving down the syringe which will show that the infusion is working. If you have any concerns that the infusion is not going as expected, contact your doctor or nurse or homecare provider.

The infusion should be completed within a maximum of 2 hours after withdrawing Aspavali from the vial, or a new syringe should be prepared, using a new vial of Aspavali. If a new syringe needs to be prepared because it has been more than 2 hours, contact your doctor or nurse, or homecare provider, to let them know.

Step 9: Complete infusion



- If after about 30 minutes from when the infusion was started the syringe looks empty, open the pump to check if the infusion is finished. The syringe will be empty when the infusion is finished.



- Once the infusion is finished, remove the empty syringe by gently pushing it back to disengage the nose. Lift the syringe up and out of the pump.



- Get some gauze and tape ready in case needed.
- One at a time, hold each needle in place and gently peel back any adhesive or dressing around it.
- Remove the needles in a straight motion, in the opposite direction you inserted them. Close the wings over the needle and snap shut.
- Remove the needles and their tubing from the syringe and throw away into the sharps container.
- If you need to, cover the infusion sites with gauze and tape.

Step 10: Record infusion



- Record your treatment as directed by your doctor or nurse.

Step 11: Clean up

- Throw away all used disposable supplies and the empty vial, as recommended by your doctor or nurse.
- If you need to clean the infusion pump, wipe the outside surface with warm water and soap. Wipe again with clean water to rinse. Read the manufacturer's instructions for more information on cleaning the infusion pump.
- Store the infusion pump in a cool, dry place.

Remember to always talk to your doctor or nurse about any questions or concerns about treatment

Recording your batch number

It's good practice to take a note of the batch number shown on your vial. You can use the template below to record the date and batch number for each infusion.

[illegible][illegible]

Tips for your infusion

To best prepare yourself for your infusion, you may wish to consider the following tips:

Stay hydrated

It is important to be hydrated ahead of your infusion.

Choose a suitable environment

Choose somewhere free from distractions, where you will be comfortable. Make sure you choose somewhere with a surface you can easily clean.

Find a dark background

Finding something dark in colour to set up your infusion on can help you with the step of preparing the needle line. The dark coloured background can make it easier to see the liquid moving down the needle line, allowing you to more precisely control the flow and stop when the liquid is 2cm away from the needle.

Set yourself up comfortably

Set yourself up somewhere comfortable in a relaxed position so that you are not squashing your infusion sites.

Avoid strenuous activity

It is suggested to avoid strenuous exercise immediately after an infusion.

Frequently Asked Questions

How should Aspaveli be stored?

Aspaveli can be used until the expiration date that is printed on the carton. Do not use the vial if the expiration date has passed.

Aspaveli should be stored in the refrigerator. It should be kept between 2°C to 8°C (35.6°F to 46.4°F), and kept in the original carton to protect it from light. It may be helpful to keep a thermometer in your refrigerator. Do not allow Aspaveli to freeze.

Do not store Aspaveli at the back of the refrigerator to avoid freezing. Store Aspaveli in the middle of the refrigerator on one of the shelves, not in the door, to minimise changes in temperature. Do not use Aspaveli if the liquid in the vial is frozen. If there is a power cut, or your refrigerator breaks, contact your homecare provider.

If you have any questions on storage of Aspaveli, contact your homecare provider or your doctor or nurse.

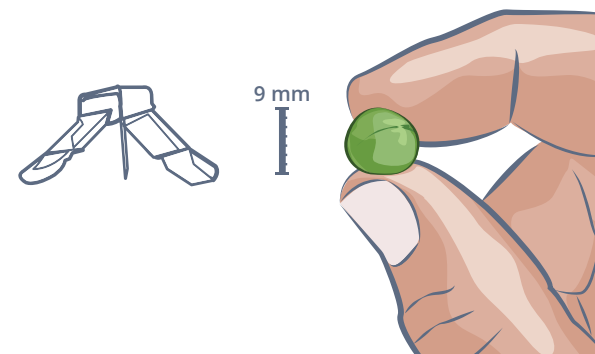
What should I do if my vials of Aspaveli freeze?

Aspaveli should not be frozen. Do not use Aspaveli if the contents of the vial become frozen at any stage. To avoid freezing, do not store Aspaveli at the back of the refrigerator.

If a vial does become frozen, contact your homecare provider for advice.

How big is the needle that I will use to administer Aspaveli?

Typically 26-gauge needles are used for the subcutaneous infusion. The needle is typically around 9 mm long, which is only about the length of a pea.



Will there be any discomfort from a subcutaneous infusion?

Subcutaneous infusions are considered to cause less discomfort than intravenous infusions.

‘Subcutaneous’ means the needle goes just under the skin. Some people may experience some discomfort at the needle site or during the infusion, however, it is mostly tolerable.

Frequently Asked Questions...continued

What should I do if I experience discomfort during the infusion or see a mark or rash on my skin at my infusion site?

If you experience any discomfort or are concerned during your infusion, contact and report it to your doctor, nurse or homecare provider. The Aspaveli Patient Information Leaflet explains the expected side effects for Aspaveli.

You should report side effects by contacting your doctor, nurse or pharmacist or you can report side effects directly via the Yellow Card Scheme at www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store. You can also report side effects to Sobi, please email drugsafety@sobi.com or call +44 (0) 800 111 4754.

What if my infusion is taking less or more time than expected?

The approximate infusion time, if you are using two infusion sites, is 30 minutes. If you are using one infusion site, it is 60 minutes. These are approximate infusion times and may be different for each person, so do not worry if your infusion is a bit faster or slower. If you have any questions about your infusion time contact your homecare provider or your doctor or nurse.

What should I do if I miss an infusion of Aspaveli?

If you miss a dose of Aspaveli, it should be taken as soon as possible, and you should let your doctor or nurse know. You should then take the next dose as regularly planned.

It is recommended that you keep a record of the doses you administer and be sure to take the record with you each time you visit your doctor or nurse because they may ask to see it.

Do I have to administer each infusion at the same time of day?

You may find it helpful to take your infusion at the same time of day to establish a routine, however it is not a problem if you change the time that you administer your infusion as long as it is on the same day as your regular schedule.

What should I do if I break a vial of Aspaveli or any of my infusion supplies, or if there are problems with my infusion pump?

If you break a vial of Aspaveli or any of your infusion supplies or have problems with or questions about your infusion pump, contact and report it to your doctor, nurse or homecare provider to let them know.

You should report product complaints (quality issues or defects with the product, pump and infusion supplies) by contacting your doctor, nurse or homecare provider. You can also report product complaints to Sobi, please email medical.info.uk@sobi.com or call +44 (0) 800 111 4754.

What you need to know before starting your treatment with Aspaveli

Risk of serious infections

The use of pegcetacoplan targets the complement system, which is part of the body's defences against infection. As such, the use of this medicine increases your risk of infections, including those caused by the so-called 'encapsulated bacteria', such as *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Haemophilus influenzae*. These are severe bacterial infections affecting your nose, throat and lungs or the linings of the brain and can spread throughout the blood and body. Serious bacterial infections may quickly become life-threatening and cause death if not recognised and treated early.

Call your doctor or nurse or seek emergency medical help right away if you have any of these signs and symptoms of a serious infection:

- Headache and a fever
- Fever and a rash
- Fever with or without shivers or chills
- Shortness of breath
- High heart rate
- Clammy skin
- Headache with a stiff neck or stiff back
- Headache with nausea (feeling sick) or vomiting
- Eyes sensitive to light
- Muscle aches with flu-like symptoms
- Confusion
- Extreme pain or discomfort

Risk of allergic reactions

Allergic reactions may occur in some patients receiving treatment with pegcetacoplan.

Immediately stop pegcetacoplan infusion if you develop any of these signs and symptoms of an allergic reaction and seek immediate medical attention:

- Difficulty breathing
- Chest pain or chest tightness
- Feeling dizzy/faint
- Severe itching of the skin or raised lumps on the skin
- Swelling of the face, lips, tongue and/or throat, which may cause difficulty in swallowing or collapse

Prophylactic vaccinations or antibiotic treatment

Vaccines against bacteria lower the risk of getting serious infections. However, vaccines do not prevent all serious infections.

Your doctor will ensure that you receive vaccination against the bacteria *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Haemophilus influenzae* if you have not had these vaccines in the past.

If you have had these vaccines in the past, you might still need additional vaccinations before starting treatment with pegcetacoplan. Your doctor will advise if you need additional vaccinations.

These vaccinations should be given at least 2 weeks before beginning pegcetacoplan therapy.

If you cannot be vaccinated 2 weeks before beginning pegcetacoplan therapy, your doctor will prescribe antibiotics (medications to treat bacterial infections) to reduce the risk of infection, for 2 weeks after you have been vaccinated. Pegcetacoplan will only be supplied if your doctor or pharmacist submits proof that you are receiving the required vaccinations or antibiotics.

Your doctor or pharmacist will receive annual vaccination reminders and will contact you in case you need revaccination. It is important that your vaccinations are up to date, so speak with your doctor or nurse about this. You should also be aware that vaccines will reduce the risk of serious infections, but do not prevent all serious infections.

Risk of destruction of red blood cells (haemolysis) after drug discontinuation

- Do not stop treatment with pegcetacoplan without discussing this beforehand with your doctor or nurse.
- It is very important to make sure that you do not miss or postpone any scheduled treatments. If you stop taking the medicine suddenly, you may be at risk of making your symptoms worse.
- If pegcetacoplan treatment is stopped completely, postponed, or if treatments are missed, there is a risk that haemolysis could occur. Haemolysis is when red blood cells, which carry oxygen through your body, break apart. Haemolysis is connected to various symptoms of PNH, such as:

- Tiredness (fatigue)
- Dark urine (haemoglobinuria)
- Tummy (abdominal) pain
- Breathlessness
- Formation of blood clots (thrombosis)
- Difficulty in swallowing
- Erectile dysfunction

- Seek immediate medical attention if you notice any signs or symptoms of haemolysis.

Reporting side effects

Reporting side effects of your treatment is important as it allows collection of more information about the safety of Aspaveli. If you experience any side effects (this also includes any possible side effects not listed in the Patient Information Leaflet), in particular serious infections with encapsulated bacteria, severe hypersensitivity reactions, or haemolysis after drug discontinuation, inform your doctor, nurse or pharmacist.

You can also report side effects directly via the Yellow Card Scheme at www.mhra.gov.uk/yellowcard or search for MHRA Yellow Card in the Google Play or Apple App Store. You can also report side effects to Sobi, please email drugsafety@sobi.com or call +44 (0) 800 111 4754.

By reporting side effects, you can help provide more information on the safety of this medicine.

Further support available

PNH Support

PNH Support is a registered charity (No 1161518) run by PNH patients and carers as a peer-to-peer support organisation for people living with PNH (and their carers and family members) in England, Wales and Northern Ireland. Our activities include: facilitating PNH patients and carers to share their experiences and support one another; advocating on behalf of PNH patients and carers with stakeholders; and providing or signposting to further information.

How to get in touch:

Email: contact@pnhuk.org

PNH Scotland

PNH Scotland is a charity dedicated to supporting anyone affected by PNH. Our main aim is to raise awareness of the condition and advance the education of patients, their families and interested health professionals. In particular, we aim to ensure all patients receive access to the treatment they require, provide up to date information and act as a point of contact for anyone needing help and advice.

How to get in touch:

Email: info@pnhscotland.org.uk

Tel: +44 (0) 7967 118762

Notes

This image shows a single sheet of white paper with horizontal blue ruling lines. The lines are evenly spaced and run across the width of the page. There are no margins, text, or other markings on the paper.

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The FreedomEdge® Syringe Infusion System, High-Flo Subcutaneous Safety Needle Sets™ and Precision Flow Rate Tubing™ are trademarks of KORU Medical Systems and are compliant with Medical Device Directive 93/42/EEC.

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NP-30393
December 2023

