

MEDICATION GUIDE

EPYSQLI® (eh-pis'-klee)

(eculizumab-aagh)

injection, for intravenous use

What is the most important information I should know about EPYSQLI?

EPYSQLI is a medicine that affects your immune system. EPYSQLI may lower the ability of your immune system to fight infections.

- **EPYSQLI increases your chance of getting serious meningococcal infections caused by *Neisseria meningitidis* bacteria. Meningococcal infections may quickly become life-threatening or cause death if not recognized and treated early.**
 - You must complete or update your meningococcal vaccines at least 2 weeks before your first dose of EPYSQLI.
 - If you have not completed your meningococcal vaccines and EPYSQLI must be started right away, you should receive the required vaccine(s) as soon as possible.
 - If you have not been vaccinated and EPYSQLI must be started right away, you should also receive antibiotics to take for as long as your healthcare provider tells you.
 - If you had a meningococcal vaccine in the past, you might need additional vaccines before starting EPYSQLI. Your healthcare provider will decide if you need additional meningococcal vaccines.
 - Meningococcal vaccines do not prevent all meningococcal infections. **Call your healthcare provider or get emergency medical care right away if you get any of these signs and symptoms of a serious meningococcal infection:**
 - fever
 - fever with high heart rate
 - headache and fever
 - confusion
 - muscle aches with flu-like symptoms
 - fever and a rash
 - headache with nausea or vomiting
 - headache with a stiff neck or stiff back
 - eyes sensitive to light

Your healthcare provider will give you a Patient Safety Card about the risk of serious meningococcal infection.

Carry it with you at all times during treatment and for 3 months after your last dose of EPYSQLI. Your risk of meningococcal infection may continue for several weeks after your last dose of EPYSQLI. It is important to show this card to any healthcare provider who treats you. This will help them diagnose and treat you quickly.

EPYSQLI is only available through a program called the EPYSQLI Risk Evaluation and Mitigation Strategy (REMS). Before you can receive EPYSQLI, your healthcare provider must:

- enroll in the EPYSQLI REMS program
- counsel you about the risk of serious meningococcal infections
- give you information about the signs and symptoms of serious meningococcal infection
- make sure that you are vaccinated against serious infections caused by meningococcal bacteria and that you receive antibiotics if you need to start EPYSQLI right away and you are not up to date on your vaccines
- give you a **Patient Safety Card** about your risk of meningococcal infection, as discussed above

EPYSQLI may also increase the risk of other types of serious infections caused by encapsulated bacteria, including *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Neisseria gonorrhoeae*.

- If your child is treated with EPYSQLI, your child should receive vaccines against *Streptococcus pneumoniae* and *Haemophilus influenzae type b* (Hib).
- Certain people may be at risk of serious infections with gonorrhea. Talk to your healthcare provider about whether you are at risk for gonorrhea infection, about gonorrhea prevention, and regular testing.
- Certain fungal infections (aspergillus) may also happen if you take EPYSQLI and have a weak immune system or a low white blood cell count.

For more information about side effects, see “**What are the possible side effects of EPYSQLI?**”

What is EPYSQLI?

EPYSQLI is a prescription medicine used to treat:

- people with paroxysmal nocturnal hemoglobinuria (PNH).
- people with atypical hemolytic uremic syndrome (aHUS).
EPYSQLI is not for use in treating people with Shiga toxin E. coli related hemolytic uremic syndrome (STEC- HUS).
- adults with generalized myasthenia gravis (gMG) who are anti-acetylcholine receptor (AChR) antibody positive.

It is not known if EPYSQLI is safe and effective in children with PNH or gMG.

Who should not receive EPYSQLI?

Do not receive EPYSQLI if you have a serious meningococcal infection when you are starting EPYSQLI treatment.

Before you receive EPYSQLI, tell your healthcare provider about all of your medical conditions, including if you:

- have an infection or fever.
- are pregnant or plan to become pregnant. It is not known if EPYSQLI will harm your unborn baby.
- are breastfeeding or plan to breastfeed. It is not known if EPYSQLI passes into your breast milk.

Tell your healthcare provider about all the medicines you take, including prescription and over-the-counter medicines, vitamins, and herbal supplements. EPYSQLI and other medicines can affect each other causing side effects.

Know the medications you take and the vaccines you receive. Keep a list of them to show your healthcare provider and pharmacist when you get a new medicine.

How should I receive EPYSQLI?

- Your healthcare provider will give you EPYSQLI into your vein through an intravenous (IV) line usually over 35 minutes in adults and 1 to 4 hours in children.
- Adults will usually receive an EPYSQLI infusion:
 - weekly for 5 weeks, then
 - every 2 weeks.
- Children less than 18 years of age, your healthcare provider will decide how often you will receive EPYSQLI depending on your age and body weight.
- After each infusion, you should be monitored for at least 1 hour for infusion-related reactions. See **“What are the possible side effects of EPYSQLI?”** If you have an infusion-related reaction during your EPYSQLI infusion, your healthcare provider may decide to give EPYSQLI more slowly or stop your infusion.
- If you miss an EPYSQLI infusion, call your healthcare provider right away.
- **If you have PNH, your healthcare provider will need to monitor you closely for at least 8 weeks after stopping EPYSQLI. Stopping treatment with EPYSQLI may cause breakdown of your red blood cells due to PNH.**

Symptoms or problems that can happen due to red blood cell breakdown include:

- drop in the number of your red blood cell count
- drop in your platelet counts
- confusion
- kidney problems
- blood clots
- difficulty breathing
- chest pain
- **If you have aHUS, your healthcare provider will need to monitor you closely for at least 12 weeks after stopping EPYSQLI for signs of worsening aHUS symptoms or problems related to abnormal clotting (thrombotic microangiopathy).**

Symptoms or problems that can happen with abnormal clotting may include:

- stroke
- confusion
- seizure
- chest pain (angina)
- difficulty breathing
- kidney problems
- swelling in arms or legs
- a drop in your platelet count

What are the possible side effects of EPYSQLI?

EPYSQLI can cause serious side effects including:

- See **“What is the most important information I should know about EPYSQLI?”**
- **Serious infusion-related reactions.** Serious infusion-related reactions can happen during your EPYSQLI infusion. Tell your healthcare provider or nurse right away if you get any of these symptoms during your EPYSQLI infusion:
 - chest pain
 - trouble breathing or shortness of breath
 - swelling of your face, tongue, or throat
 - feel faint or pass out

If you have an infusion-related reaction to EPYSQLI, your healthcare provider may need to infuse EPYSQLI more slowly, or stop EPYSQLI. See **“How will I receive EPYSQLI?”**

The most common side effects in people with PNH treated with EPYSQLI include:

- headache
- back pain
- pain or swelling of your nose or throat (nasopharyngitis)
- nausea

The most common side effects in people with aHUS treated with EPYSQLI include:

- headache
- stomach-area (abdominal) pain
- low red blood cell (anemia)
- nausea
- diarrhea
- vomiting
- cough
- urinary tract infections
- high blood pressure (hypertension)
- pain or swelling of your nose or throat
- swelling of legs or feet (peripheral edema)
- fever
- common cold (upper respiratory infection)
- (nasopharyngitis)

The most common side effects in people with gMG treated with EPYSQLI include:

- muscle and joint (musculoskeletal) pain

Tell your healthcare provider about any side effect that bothers you or that does not go away. These are not all the possible side effects of EPYSQLI.

Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.

General information about the safe and effective use of EPYSQLI.

Medicines are sometimes prescribed for purposes other than those listed in a Medication Guide. You can ask your pharmacist or healthcare provider for information about EPYSQLI that is written for health professionals.

What are the ingredients in EPYSQLI?

Active ingredient: eculizumab-aagh

Inactive ingredients: dibasic sodium phosphate, monobasic sodium phosphate, polysorbate 80 (vegetable origin), trehalose, and Water for Injection.

Manufactured by Samsung Bioepis Co., Ltd., 76, Songdogoyuk-ro, Yeonsu-gu, Incheon 21987 Republic of Korea. US License Number 2046

This Medication Guide has been approved by the U.S. Food and Drug Administration.

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