

HEALTHCARE PROFESSIONAL RESOURCE AND TRAINING GUIDE **ALL ABOUT HAEGARDA®**

This Resource Guide is intended to be a tool to educate Health Care Professionals on HAEGARDA® Home Infusion, and not for patient training.

For patient training, see the HAEGARDA® Patient Resource Binder, available through CSL Behring Canada.

PATIENTS SELF-ADMINISTER HAEGARDA®

LEARNING OBJECTIVES

After completing this chapter, you will be able to:

- ✓ Educate patients on the Patient Support Program (CSL Behring PLUS+).
- ✓ Explain clinical characteristics of HAE and how to treat patients.
- ✓ Educate patients about HAEGARDA®.
- ✓ Share important safety information, warning and precautions when using HAEGARDA® with patients.
- ✓ Develop competency in training patients to safely self-administer HAEGARDA® for routine prevention of HAE attacks.



HAE, hereditary angioedema; C1-INH, C1 esterase inhibitor



ENROLLING YOUR PATIENT IN CSL BEHRING PLUS+

CSL Behring PLUS+:

- Simple and easy enrolment
- Comprehensive training for self-administration of HAEGARDA® by a qualified nurse educator
- Resources and educational material on subcutaneous self-administration
- Additional supplies and reimbursement support when needed
- Follow-up to maintain patient competency level for subcutaneous self-administration



HELPING PATIENTS SELF-ADMINISTER HAEGARDA®

CSL BEHRING PLUS+

Physician faxes Patient Enrolment form to CSL Behring PLUS+

1-888-490-4106

Patient can also call 1-888-490-4105 directly provided s/he has valid prescription

Physician coordinates with the blood bank to ensure that HAEGARDA® is released to the patient for self-administration

It is recommended that the patient has product on hand before the training is conducted

Nurse educator contacts the patient and sets up a training date and logistics

A report is sent to the treating physician each time the patient is trained

Blood bank receives a report when patient completes training

The patient is trained as many times as needed to ensure that s/he is competent to self-administer HAEGARDA®.

Supplies provided as per the nurse educator's recommendations

Nurse case manager follows up with the patient on a regular basis to ensure continued patient competency for SC self-administration

The nurse sends a summary note to the physician when the patient is contacted

Follow-up dates are flexible for the patient's convenience

CSL Behring

PLUS+
PATIENT SUPPORT PROGRAM



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 **HAEGARDA®**
C1 Esterase Inhibitor
Subcutaneous (Human)

CSL BEHRING PLUS+

Follow-up dates are flexible for the patient's convenience

Pre-training call

Week 1 follow-up

Week 3 follow-up

Semi-annual
follow-ups

Month 6 follow-up

Month 3 follow-up

THE HAEGARDA® PACKAGE CONTENTS

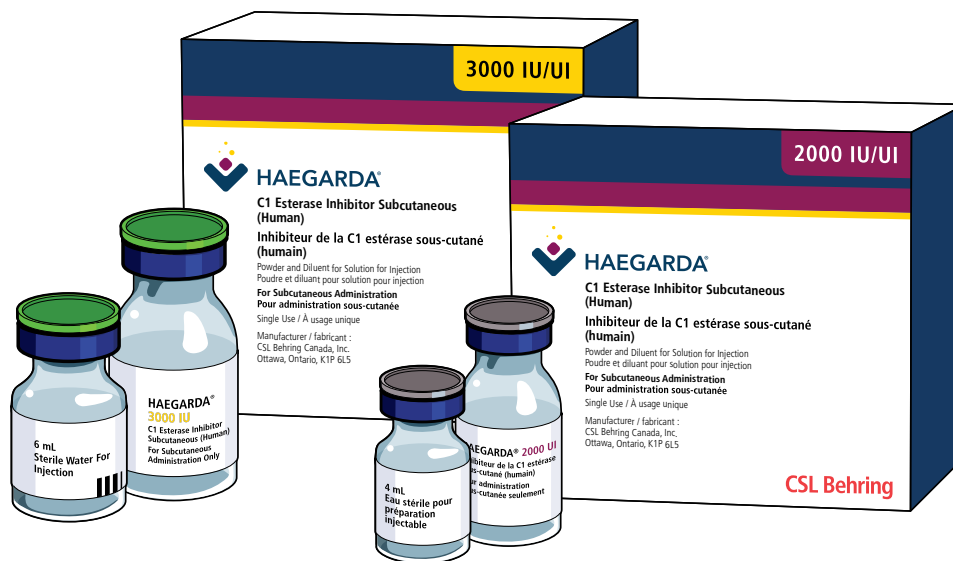
The product package includes:

- 1 vial with HAEGARDA® powder
- 1 vial of diluent (Sterile Water for Injection)
- 1 Mix2Vial® transfer device for reconstitution
- 1 inner carton

The inner carton contains:

- 1 syringe (10 mL) for withdrawal
- 1 subcutaneous infusion set
- 1 hypodermic needle

The components used in the packaging for HAEGARDA® are latex-free



For additional supplies or questions please contact CSL Behring PLUS+ at:
1-888-490-4105 or cslbehringplus@innomar-strategies.com



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WHAT YOUR PATIENT NEEDS TO KNOW ABOUT THE HAEGARDA® ENROLMENT PROCESS

Patients should expect an enrolment call from CSL Behring PLUS+ that will last approximately 30 minutes (depending on the questions from the patient).

This is what happens during a typical call and what the patient should expect:

- The program will need to confirm the details of the patient's demographics, prescription, physician name and blood bank to ensure the information is accurate.
- The patient will then be given details on training expectations, responsibilities and how to store the product.
- The training nurse will coordinate a training date with the patient, and will provide as many training sessions as needed until the patient is comfortable with self-administration.
- The training can be done either at a private clinic or at the patient's home, depending on their preference and/or their healthcare professional's orders on the enrolment form.
- The patient can call the program number (1-888-490-4105) for any questions and support.
- The program will follow up with the patients at regular intervals starting at 1 week, 1 month, 3 months then every 6 months going forward. These touch points are flexible, depending on patient's needs.

ELEMENTS FOR SUCCESSFUL TRAINING

The Nurse as an Essential Element in HAE Patient Care

- Educates, advocates, manages patients and healthcare team
- Encourages patients to take ownership of therapy



HAEGARDA® SELF-ADMINISTRATION GUIDE

STEP-BY-STEP INSTRUCTIONS FOR PREPARING AND ADMINISTERING HAEGARDA®

Use this guide in conjunction with PART III (Consumer Information) of the Product Monograph and training provided by a healthcare professional (HCP).

Do not attempt to self-administer unless you have been trained by a healthcare professional.



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IMPORTANT SAFETY INFORMATION

Home treatment/Self-administration:

You should prepare the prescribed dose of HAEGARDA® for self-administration as directed by your healthcare professional.

- **Do not attempt to self-administer unless you have been taught how by your healthcare professional.**
- Your healthcare professional will prescribe the dose that you should administer, which is based on your body weight.
- Talk to your healthcare professional before traveling to make sure you have an adequate supply of HAEGARDA®.
- Use a new needle for each HAEGARDA® injection.

Reconstitution and Administration:

HAEGARDA® is intended for self-administration by subcutaneous injection only. The patient or caregiver should be trained on how to administer HAEGARDA® as needed.

Training Requirements

- It is important to help manage patient expectations on HAEGARDA® therapy so that they are better prepared when variabilities arise, and less likely to be non-adherent to therapy.
- Explain benefits/risks of HAEGARDA® therapy/reasons for choice.
- Set expectations:
 - how long an infusion takes
 - necessary equipment
 - transition/adjustment period to home infusion therapy



IMPORTANT SAFETY INFORMATION

Side effects and what to do about them

These are not all the possible side effects you may feel when taking HAEGARDA®. If you experience any side effects not listed here, contact your healthcare professional.

Allergic reactions may occur with HAEGARDA®

Talk to your healthcare professional right away if you have any of the following symptoms after using HAEGARDA®:

- wheezing
- difficulty breathing
- chest tightness
- turning blue (look at lips and gums)
- fast heartbeat
- swelling of the face
- rash or hives



IMPORTANT SAFETY INFORMATION

Signs of a blood clot include:

- pain and/or swelling of an arm or leg with warmth over the affected area
- discoloration of an arm or leg
- unexplained shortness of breath
- chest pain or discomfort that worsens on deep breathing
- unexplained rapid pulse
- numbness or weakness on one side of the body

The most common side effects with HAEGARDA® are injection site reactions (pain, redness, swelling), hypersensitivity (itching and rash), nasopharyngitis (runny or stuffy nose, sneezing, watery eyes) and dizziness.

If you have a troublesome symptom or side effect that is not listed here or becomes bad enough to interfere with your daily activities, talk to your healthcare professional.



WARNINGS AND PRECAUTIONS

Do not use HAEGARDA® if:

You have experienced life-threatening immediate hypersensitivity reactions, including anaphylaxis, to the product.

To help avoid side effects and ensure proper use, talk to your healthcare professional before you take HAEGARDA®. Talk about any concerns, health conditions or problems you may have, including the following:

- You are pregnant or planning to become pregnant. The effect of HAEGARDA® on your unborn baby is not known.
- You are breastfeeding or plan to breastfeed. It is not known if HAEGARDA® passes into your breast milk or if it can affect your baby.
- You have a history of blood clotting problems. Blood clots have occurred in patients receiving HAEGARDA®. Very high doses of C1-INH could increase the risk of blood clots. Tell your healthcare professional if you have a history of heart or blood vessel disease, stroke, blood clots, or have thick blood, an indwelling catheter/access device in one of your veins, or have been immobile for some time. These factors may increase your risk of having a blood clot after using HAEGARDA®. Also, tell your healthcare professional what drugs you are using, as some drugs, such as birth control pills or certain androgens, may increase your risk of developing a blood clot.



WARNINGS AND PRECAUTIONS

Tell your healthcare professional about all the medicines you take, including any drugs, vitamins, minerals, natural supplements or alternative medicines.

Products made from human plasma may contain infectious agents such as viruses and, theoretically, the agent responsible for the Creutzfeldt-Jakob disease (CJD).

Appropriate vaccination (hepatitis A and B) should be generally considered for subjects in regular/repeated receipt of human plasma-derived products.



MY HEALTHCARE TEAM

| CONTACT | NAME | CONTACT NUMBER |
|-------------------------|------|----------------|
| Clinic Nurse | | |
| Doctor | | |
| Emergency Department | | |
| Blood Bank | | |
| Other Relevant Contacts | | |

Getting your HAEGARDA®

- HAEGARDA® is a plasma-derived C1-esterase inhibitor which is typically distributed by the Blood Bank.
- Instructions may differ in each center as to where and how to pick up your HAEGARDA® supply.
Please follow the instructions outlined by your healthcare professional to obtain your HAEGARDA® supply.



ABOUT THIS GUIDE

HAEGARDA® is an injectable medicine used to prevent swelling and/or painful attacks in adults and adolescents with Hereditary Angioedema (HAE). HAEGARDA® should not be used to treat an acute HAE attack. In the event of an acute attack, seek medical attention.

This self-administration guide can help you through the reconstitution and injection process, especially when you are first learning. It should be used in conjunction with training provided by a healthcare provider.

This guide is divided into 3 sections:

- Preparation and handling
- Reconstitution
- Self-administration



SETTING UP A TREATMENT SCHEDULE

HAEGARDA® should be taken as prescribed by your healthcare professional. HAEGARDA® is administered by subcutaneous self-administration, so you can choose the time and place that works best for you.

Here are a few tips to help make taking HAEGARDA® part of your routine

- See the following step-by-step instructions for the reconstitution and administration of HAEGARDA®. The following steps are general guidelines for using HAEGARDA®. If you are unsure of the steps, please contact your healthcare professional before using.
- Use a new needle for each HAEGARDA® injection.
- If you miss a dose of HAEGARDA®, proceed with your next dose immediately and continue at regular intervals as advised by your healthcare professional.

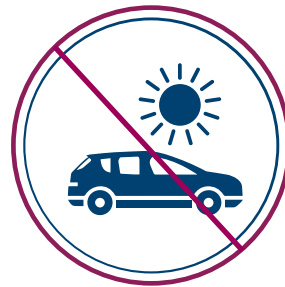
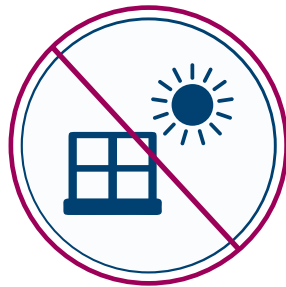
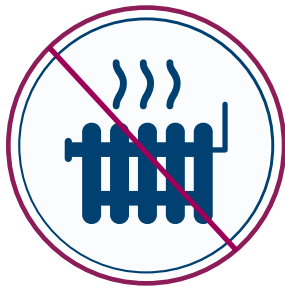
Do not take a double dose to make up for a forgotten dose.



PREPARATION AND HANDLING: STORING HAEGARDA®

- Store HAEGARDA® in the refrigerator or at room temperature (at +2°C to +30°C). Do not use the product after the expiration date.
- Keep HAEGARDA® in its original carton until ready to use. Do not freeze. Protect from light.
- Avoid extreme temperatures (outside the range indicated above).

See some examples of extreme temperatures:



Tips for Traveling with HAEGARDA®

- Take an adequate quantity of HAEGARDA® product and supplies (for example: Syringes, Mix2Vial®, etc.) to cover treatment during your traveling period. Discuss with your healthcare professional how many doses you should always keep with you.
- The product must be kept at +2°C to +30°C:
 - It must be protected from temperature extremes
- Take a copy of your prescription for HAEGARDA® with you (if required).
- Also take any other documentation you may need such as:
 - Various letters
 - Wallet card
 - Infusion diary/log sheets
- Ask your healthcare professional for a travel letter.
- Always keep your HAEGARDA® in your carry-on bag, not in your checked luggage.



PREPARATION AND HANDLING

>>STEP 1:

Assemble the following supplies

- HAEGARDA® and diluent vials (**Ensure that the HAEGARDA® and the diluent are at room temperature**). Check the expiration date on the product vial label. Do not use the vial if it is beyond the expiration date.
- Mix2Vial®
- SC infusion set or hypodermic needle
- Sterile syringe
- Alcohol or disinfectant wipes
- Sharp/biohazardous container
- Treatment diary/log book
- Gloves (if recommended by your healthcare professional)

Use a new needle for every HAEGARDA® injection. Do not reuse or share needles with other people.

HCP Notes:

- Assemble Supplies.
- Instruct the patient to assemble all the supplies needed for treatment.
- The patient should also have extra supplies on hand in case of contamination.

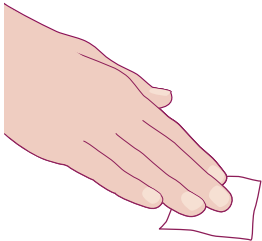


PREPARATION AND HANDLING

>>STEP 2:

Clean surface

Thoroughly clean a table or other flat surface using alcohol or disinfectant wipes.



>>STEP 3:

Wash hands

Thoroughly wash and dry your hands. If you have been told to wear gloves when preparing your infusion, put the gloves on.



You can use the mat included in your HAEGARDA® Welcome Kit as a surface to prepare HAEGARDA® for self-administration. Clean the mat with an antiseptic wipe before each use.

HCP Notes:

- Clean Surface
- Instruct the patient to locate a flat, non-porous surface/infusion mat and clean the area with an alcohol wipe/disinfectant.
- Thoroughly wash and dry your hands.
- Instruct the patient to thoroughly wash and dry hands.
- Check your institution's guidelines to confirm their recommended length of time and method of contamination prevention. Some examples include: aseptic technique, minimum 20 seconds, 2 "Happy Birthdays", or the use of paper towels to turn off taps, etc.
- If hands are not visibly soiled, clean your hands by rubbing them with an alcohol-based formulation (hand sanitizer containing 70% alcohol), as the preferred way for routine hand hygiene.



RECONSTITUTING AND ADMINISTERING

>>STEP 4:

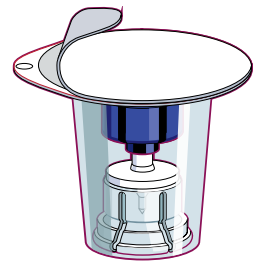
Clean stoppers

Inspect each product vial. Check expiration date. Ensure the caps are secure. Remove the flip caps from both vials (HAEGARDA® and diluent). Wipe rubber stoppers with an antiseptic wipe and allow the rubber stopper to dry.



>>STEP 5:

Open the Mix2Vial® package by peeling off the lid. Do not remove the Mix2Vial® from the blister package!



HCP Notes:

Inspection of the Vial:

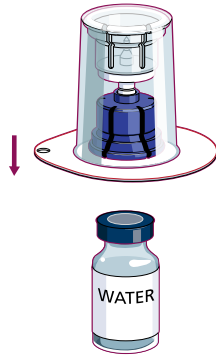
- No cracks or damage to the vials
 - Protective caps intact.
 - If the vial or protective cap is damaged, do not use
-
- Return vial to the TMS/Blood Bank.
 - When cleaning the rubber stoppers, use one antiseptic wipe per vial, ensuring aseptic technique.
 - Remove the Mix2Vial® Packaging.
 - Ensure the patient does not touch the spike in the blue top or the exposed spike in the clear end.



RECONSTITUTING AND ADMINISTERING

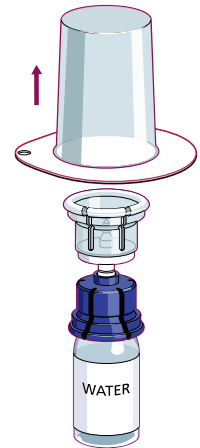
>>STEP 6:

Place the diluent vial on an even, clean surface and hold the vial tight. Take the Mix2Vial® together with the blister package and push the spike of the blue adapter end straight down through the diluent vial stopper.



>>STEP 7:

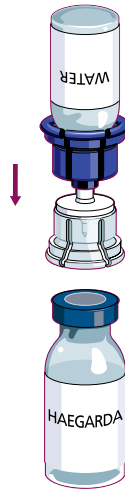
Carefully remove the blister package from the Mix2Vial® set by holding at the rim, and pulling vertically upwards. Make sure that you only pull away the blister package and not the Mix2Vial® set.



RECONSTITUTING AND ADMINISTERING

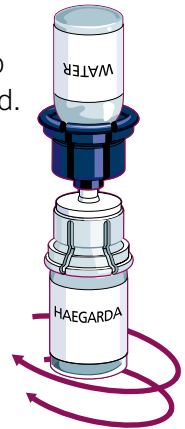
>>STEP 8:

Place the HAEGARDA® vial on an even and firm surface. Invert the diluent vial with the Mix2Vial® set attached and push the spike of the transparent adapter end straight down through the HAEGARDA® vial stopper. The diluent will automatically flow into the HAEGARDA® vial.



>>STEP 9:

With the diluent and HAEGARDA® vial still attached to the Mix2Vial® transfer set, gently swirl the HAEGARDA® vial to ensure that the powder is fully dissolved. Do not shake the vial.



HCP Notes:

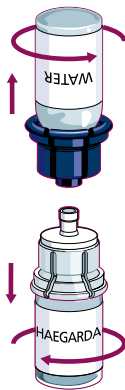
- Transfer Diluent/Sterile Water into the HAEGARDA® vial.
- The Mix2Vial® should be aligned completely vertical before piercing the rubber stopper.
- The vacuum in the HAEGARDA® vial will allow the diluent to transfer into the HAEGARDA® vial automatically. The vacuum loss may occur if the spike is inserted at an angle.
- If the fluid does not transfer automatically, instruct the patient to contact the training nurse for further instructions.
- Instruct the patient to use new vials of HAEGARDA® and diluent if the diluent does not transfer into the HAEGARDA® powder.



RECONSTITUTING AND ADMINISTERING

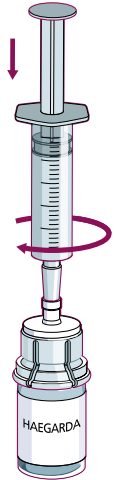
>>STEP 10:

With one hand grasp the HAEGARDA® side of the Mix2Vial® set and with the other hand grasp the diluent-side and unscrew the set carefully counter-clockwise into two pieces. Discard the diluent vial with the blue Mix2Vial® adapter attached.



>>STEP 11:

Draw air into an empty, sterile syringe. While the HAEGARDA® vial is upright, connect the syringe to the Mix2Vial®'s Luer Lock fitting by screwing clockwise. Inject air into the HAEGARDA® vial.



RECONSTITUTING AND ADMINISTERING

>>STEP 12:

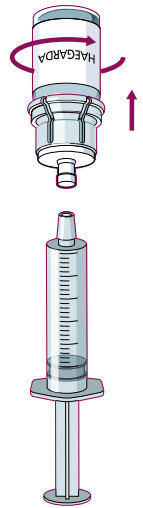
While keeping the syringe plunger pressed, turn the system upside down and draw the solution into the syringe by pulling the plunger back slowly.



>>STEP 13:

Now that the solution has been transferred into the syringe, firmly hold on to the barrel of the syringe (keeping the syringe plunger facing down) and disconnect the transparent Mix2Vial® adapter from the syringe by unscrewing counter-clockwise. The reconstituted solution should be colorless, clear and free from visible particles.

Do not use if particulate matter or discoloration is observed.



HCP Notes:

- Prepare Administration Set.
- Ensure patient has the infusion butterfly prepared to attach to the HAEGARDA®-filled syringe to avoid contamination of the syringe.



SELF-ADMINISTRATION

>>STEP 14:

Prepare injection site

- Select an area on the abdomen (stomach; Figure 1) or another subcutaneous area for the injection as discussed with a healthcare professional.
- Use a different place from your last injection.
- New injection sites should be at least 5 centimeters (2 inches) away from the place where injection was given previously.
- Never give injection in areas where the skin is itchy, swollen, painful, bruised, or red.
- Avoid giving injections in places with scars or stretch marks.
- Clean the skin at the injection site with an alcohol swab and let the skin dry (Figure 2).

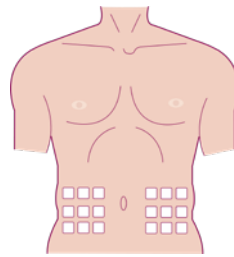


Figure 1

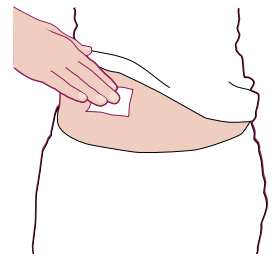


Figure 2



SELF-ADMINISTRATION

>>STEP 15:

Injection in the abdominal area or other subcutaneous injection area

As instructed by a healthcare provider:

- Attach a hypodermic needle or SC infusion set. Prime the needle or tubing as required and instructed.

Injection with hypodermic needle:

- Insert the needle into the fold of skin (Figure 3)

Injection by SC Infusion Set:

- Insert the needle into the fold of skin (Figure 4)

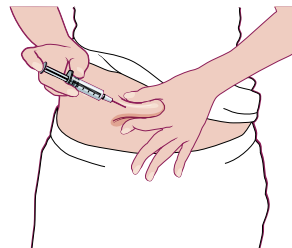


Figure 3

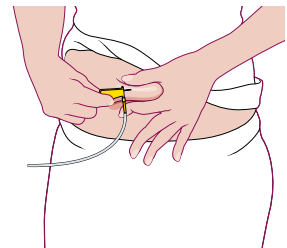


Figure 4



AUTO-ADMINISTRATION

>>STEP 16:

Clean up

- After injecting the entire amount of HAEGARDA®, remove the needle.
- Any unused medicinal product or waste material should be disposed of in accordance with local requirements.



>>STEP 17:

Record treatment

You may also use the HAERO app to track and record your treatments.

Download the HAERO app from the App store or Google play to track your attacks and treatment.



HCP Notes:

How to Dispose of a Full Sharps Container

- Ensure the lid is closed tightly.
- DO NOT dispose of a sharps container in regular garbage or recycling.
- The patient should be instructed to check with their municipality to determine their local policy regarding sharps containers.

Record the Infusion

- Discuss the importance of documentation of all information, including lot numbers.
- Documentation of attacks helps to ensure best treatment for the patient. It also tracks the product in case of recall.



KEY LEARNINGS

1

Strategies to educate patients about HAE, what HAEGARDA® is and how it should be handled.

2

Information about safety information, warnings and precautions that should be explained to patients.

3

Strategies for successfully teaching patients how to self-administer HAEGARDA®.

4

Information on how to enroll patients in CSL Behring PLUS+.



HAE, hereditary angioedema



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