ALBERTA PRECISION LABORATORIES
Leaders in Laboratory Medicine

Haegarda®
C1 Esterase Inhibitor Subcutaneous (human)

APPLICABILITY: This document applies to APL, AHS, Covenant Health, and all other health care professionals involved in the transfusion of blood components and products in Alberta.	***Other Names:*** *C1 Inhibitor* ***Company:*** *CSL Behring* ***Class:*** *Manufactured blood product, derived from human plasma*

	INTRAVENOUS			OTHER		
ROUTES	**DIRECT IV**	**IV Infusion**	**Continuous Infusion**	**SC**	**IM**	**OTHER**
Acceptable Routes*	No	Yes**	No	Yes	No	N/A

* Administration of blood components and blood products is a restricted activity. For specific conditions that apply to a profession's authorization to administer blood components and blood products, consult the applicable discipline-specific regulation under the Health Professions Act (Alberta). Health care professionals with this authorization require the applicable education, training and competency.
****** Intravenous administration is currently considered off-label use and may only occur at the direction of the Rare Blood Disorders / Angioedema Clinic physician.

DESCRIPTION:

- HAEGARDA® C1 Esterase Inhibitor Subcutaneous (human) is a purified, pasteurized, nanofiltered, lyophilized concentrate of C1 Esterase Inhibitor (C1-INH) derived from large pools of human plasma.
- Product is a white lyophilized powder.
- Reconstituted solution is colourless and clear to slightly opalescent.
- Viral inactivation steps include pasteurization, hydrophobic interaction chromatography, and nanofiltration.
- Supplied in 2000 IU and 3000 IU vials.
- Each vial is reconstituted with sterile water for injection, resulting in a concentration of 500 IU/mL.
- Also contains glycine, sodium chloride, and sodium citrate.
- Preservative-free
- Latex-free

AVAILABILITY

- Supplied by Canadian Blood Services.
- Contact your local laboratory/transfusion service regarding stock availability on site.

INDICATIONS:

- For routine prevention of angioedema attacks in adult and adolescents with hereditary angioedema (HAE).

CONTRAINDICATIONS:

- Patients who are hypersensitive (allergic) to this blood product or any ingredient in its formulation (see description of product)
- HAEGARDA® should not be used to treat an acute HAE attack.

WARNINGS:

- Hypersensitivity reactions may occur.
- Thrombotic events have occurred in patients receiving high doses of C1-INH intravenous (IV). A causal relationship with recommended subcutaneous doses has not been established.

DOSE (Refer to Product Insert):

- Dose to be determined by the most responsible health practitioner (MHRP).
- Consult with Hematologist or bleeding disorders clinic.
- For routine prevention against angioedema attacks, manufacturer recommended dose is 60 IU/kg body weight, twice weekly (every 3 or 4 days).

ADMINISTRATION:

Confirm written (signed) consent has been obtained and documented prior to requesting blood component from lab/transfusion service where possible.

Pre-Infusion:

- Ensure recent patient weight and height is on file
- Ensure pertinent labs are available as required
- Ensure any ordered premedications have been given (antihistamines, antipyretics prn).
- Perform the appropriate pre-transfusion checks per protocol.
- Report any new onset acute illness to MD/authorized prescriber prior to commencing infusion.

Access:

- Product should be given by subcutaneous injection. Intravenous administration may be performed at the direction of a hematologist or the bleeding disorders or angioedema clinic physician.
- Suggested infusion site is the abdominal area or other subcutaneous injection sites.

Reconstitution Supplies:

- HAEGARDA® Product (lyophilized powder)
- Sterile water for injection (4 mL for 2000 IU vial or 6mL for 3000 IU vial) (included with product)
- Mix2Vial™ filter transfer device (included with product)
- Disinfectant wipes (not included with product)

Administration Supplies:

- Disinfectant wipes (not included with product)
- 10 mL syringe (included with product)
- Subcutaneous infusion set or hypodermic needle (included with product)

Reconstitution:

- Bring product to room temperature prior to reconstitution.
- See *Mix2Vial® Reconstitution Instructions.*
- Do not refrigerate after reconstitution.
- Store reconstituted product in the vial (not a syringe).

Administration:

- Reconstituted solution must be used within 8 hours of reconstitution.
- Do not mix with any other medications or solutions.
- Do not use solutions that are cloudy, contain particulates, or are discoloured.
- Administer subcutaneously in the abdominal area or other subcutaneous injection sites.
- Use a different injection site, at least 5 centimeters from the site used for the previous injection.
- **Administration rate:**
  - Administration rate should be specified by the MRHP after patient assessment.
  - Infuse at a rate tolerated by the patient.

NURSING IMPLICATIONS:

Patient Monitoring:

- Vital Signs: Pre-administration**,** on completion of dose, and as patient condition requires.

Patients receiving blood product transfusions must be observed closely for signs of any unexpected or untoward reactions. These reactions may occur during or after the infusion of blood or blood products. For follow up instructions to a transfusion reaction, go to: www.albertahealthservices.ca/lab/page4240.aspx

Documentation:

- The transfusion documentation should be double signed to indicate infusion.
- Start and stop time of infusion and assessment of patient tolerability should also be documented in appropriate flow chart or clinical record (electronic or paper) as required.
- Provide patient notification documentation where required.

POTENTIAL HAZARDS WITH PARENTERAL ADMINISTRATION:

- Potential adverse events related to a blood product transfusion range in severity from minor with no sequelae to life-threatening.
- All adverse events occurring during a transfusion should be evaluated to determine whether or not the transfusion can be safely continued/restarted.
- All adverse events suspected to be related to a product transfusion (whether during or after a transfusion) must be reported to your local transfusion service.
- The most commonly reported adverse reaction in patients receiving HAEGARDA® were injection site reaction, hypersensitivity, nasopharyngitis, and dizziness.

Side Effects - Flushing - Headache - Nausea - Itching and redness at the venipuncture site	→	**Action:** Slow rate of infusion
Potential Allergic Reaction - Stuffy nose - Hives/severe itching - Cough - Chest pain - Wheezing - Facial swelling - Fainting	→	**Action:** **STOP** infusion **IMMEDIATELY** and contact physician

STORAGE & STABILITY:

- Store at 2-30°C until expiry (up to 36 months from manufacture).
- Do not freeze.
- Protect from light.
- Do not use expired product.

CONTACT INFORMATION:

Approved By: APL Transfusion Medicine Discipline Council
For questions or comments please contact: Transfusion.SafetyTeam@albertaprecisionlabs.ca

REFERENCES:

CSL Behring Canada Inc. 3 November 2021. Haegarda® Product Monograph. Submission Control No 255230. [Accessed 10Jun22]. https://labeling.cslbehring.ca/PM/CA/HAEGARDA/EN/HAEGARDA-Product-Monograph.pdf

Betschel et al. The International / Canadian Hereditary Angioedema Guideline Allergy Asthma Clin Immunol (2019) 15:72