Cigna National Formulary Coverage Policy

Effective Date **4/1/2023**
Next Review Date **4/1/2024**

Drug Quantity Management – Per Days
Hereditary Angioedema – Haegarda

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Product Identifier(s)

132018

INSTRUCTIONS FOR USE
The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations.

National Formulary Medical Necessity

Drugs Affected

- Haegarda® (C1 esterase inhibitor [human] subcutaneous injection)

This Drug Quantity Management program has been developed to promote the safe, effective, and economic use of Haegarda. If the Drug Quantity Management rule is not met for the requested medication at the point of service, coverage will be determined by the Criteria below. All approvals are provided for 1 year in duration.

Drug Quantity Limits

Product	Package Size	Retail Maximum Quantity per 28 Days	Home Delivery Maximum Quantity per 84 Days
Haegarda® (C1 esterase inhibitor [human] SC injection)	2,000 IU single-dose vial	24 vials*	72 vials*
	3,000 IU single-dose vial	16 vials*	48 vials*

SC – Subcutaneous; * The quantity limits in this policy provide quantity sufficient for a dose of 60 IU/kg twice weekly for an individual weighing up to 100 kg, using either the 2,000 IU strength vials or the 3,000 IU strength vials. If the individual requires additional quantity based on body weight > 100 kg, exceptions are provided in criteria.

Cigna covers quantities as medically necessary when the following criteria are met:

Haegarda 2,000 IU vials:

1. If the individual weighs > 100 kg, approve the requested quantity, not to exceed 48 vials per 28 days at retail or 144 vials per 84 days at home delivery.

Haegarda 3,000 IU vials:

1. If the individual weighs > 100 kg, approve the requested quantity, not to exceed 32 vials per 28 days at retail or 96 vials per 84 days at home delivery.

Conditions Not Covered

Any other exception is considered not medically necessary.

Background

Overview

Haegarda, a human plasma-derived C1 esterase inhibitor (C1-INH), is indicated for **routine prophylaxis to prevent hereditary angioedema attacks** in adults and pediatric patients ≥ 6 years of age.[1]

Dosing

Haegarda is dosed 60 IU/kg twice weekly (once every 3 or 4 days).[1] Thus, eight doses of Haegarda are required in a 28-day period and 24 doses are required in a 84-day period.

Availability

Haegarda is available in single-dose vials of lyophilized powder containing 2,000 IU or 3,000 IU.[1] Refer to Table 1 for the number of doses required based on patient weight.

Due to the weight-based dosing of Haegarda and the vial sizes available, the maximum approvable quantity per 28 days is dependent on patient weight. The quantity limits provided in this policy provide a sufficient quantity for a patient weighing up to 100 kg using either 2,000 IU vials or 3,000 IU vials. For a patient requiring additional quantity based on body weight, quantity limits are provided in the exception criteria below.

Table 1. Vials of Haegarda Required Based on Patient Weight.

Weight (kg)	IU required per dose (rounded to nearest package size)	2,000 IU vials per dose	3,000 IU vials per dose	2,000 IU vials per 28 days	3,000 IU vials per 28 days
≤ 33 kg	2,000 IU	1	0	**8**	**0**
> 33 kg to 50 kg	3,000 IU	0	1	**0**	**8**
> 50 kg to 66 kg	4,000 IU	2	0	**16**	**0**
> 66 kg to 100 kg	6,000 IU	3*	2*	**24***	**16***
> 100 kg to 133 kg	8,000 IU	4	0	**32**	**0**
> 133 kg to 150 kg	9,000 IU	0	3	**0**	**24**
> 150 kg to 166 kg	10,000 IU	5	0	**40**	**0**
> 166 kg to 200 kg	12,000 IU	6**	4**	**48****	**32****

* Can use three 2,000 IU vials or two 3,000 IU vials to achieve the desired dose.
** Can use six 2,000 IU vials or four 4,000 IU vials to achieve to desired dose.

References

1. Haegarda® subcutaneous injection [prescribing information]. Kankakee, IL: CSL Behring; January 2022.

Revision History

Type of Revision	Summary of Changes	Approval Date
New Policy	--	01/18/2023