C1 INHIBITOR HUMAN



Included Products: CINRYZE (C1 INHIBITOR HUMAN), HAEGARDA (C1 INHIBITOR HUMAN)

Created: 07/16/2009 Revised: 03/13/2025 Reviewed: 03/13/2025 Updated: 04/01/2025

Nonformulary for outpatient benefit. PA required on medical benefit.

All	Diagnoses		
	ial Criteria: All Diagnoses	If yes	If no
1.	Does the member have a diagnosis of hereditary angioedema (HAE) confirmed by genetic testing or normal C1q lab levels with levels below the lab's normal reference range for both C4 and C1INH?	Continue to #2.	Do not approve.
2.	Does the member have a history of attacks that are considered severe with swelling of the face, throat or gastrointestinal tract that significantly interrupts usual daily activity despite short-term symptomatic treatment?	Continue to #3.	Do not approve.
3.	Has the member been evaluated for triggers of HAE attacks and is maximally managed for avoidance of those triggers (such as stress, hormonal changes, dental surgery, trauma, medications including ACE inhibitors and estrogen)?	Continue to #4.	Do not approve.
4.	Is treatment with acute, abortive therapy an option for this member (Firazyr, Berinert)?	Do not approve.	Continue to #5.
5.	Is the request for Haegarda?	Continue to #7.	Continue to #6.
6.	Review case with medical director for consideration of approval. Long-term prevention: 1000 units IV every 3-4 days. Short-term prevention: 1000 units per procedure.		
7.	Does the member weigh 100kg or less?	Continue to #9.	Continue to #8.
8.	Has the member tried and failed Cinryze IV?	Continue to #9.	Do not approve.
9.	All approvals subject to medical director review. Approvals will be limited to appropriate weight based dose every 3-4 days.		

Renewal Criteria		If yes	If no
1.	Has there been at least a 50% reduction in the number of angioedema attacks, significant improvement in the severity and duration of attacks, and clinical documentation of functional improvement?	Continue to #2.	Do not approve.
2.	Approve previous quantity for 12 months.		