



## Berinert, Cinryze

Fax completed form to: (855) 840-1678

If this is an URGENT request, please call (800) 882-4462  
(800.88.CIGNA)

PHYSICIAN INFORMATION			PATIENT INFORMATION		
* Physician Name:			*Due to privacy regulations we will not be able to respond via fax with the outcome of our review unless all asterisked (*) items on this form are completed.*		
Specialty:	* DEA, NPI or TIN:				
Office Contact Person:			* Patient Name:		
Office Phone:			* Cigna ID:		* Date of Birth:
Office Fax:			* Patient Street Address:		
Office Street Address:			City:	State:	Zip:
City:	State:	Zip:	Patient Phone:		
<b>Urgency:</b> <input type="checkbox"/> Standard <input type="checkbox"/> Urgent (In checking this box, I attest to the fact that applying the standard review time frame may seriously jeopardize the customer's life, health, or ability to regain maximum function)					
<b>Medication requested:</b> <input type="checkbox"/> Berinert 500 unit kit <input type="checkbox"/> Cinryze 500 unit vial  Directions for use: Quantity: Duration of therapy: J-Code: (for Berinert) What is your patient's current weight (kg)? ICD10:  Is this a new start or continuation of therapy with the requested drug <input type="checkbox"/> new start of therapy <input type="checkbox"/> continuation of therapy start date:					
<b>Where will this medication be obtained?</b> <input type="checkbox"/> Accredo Specialty Pharmacy** <input type="checkbox"/> Hospital Outpatient <input type="checkbox"/> Retail pharmacy <input type="checkbox"/> Other (please specify): <div style="text-align: right;"><input type="checkbox"/> Home Health / Home Infusion vendor <input type="checkbox"/> Physician's office stock (billing on a medical claim form) **Cigna's nationally preferred specialty pharmacy</div>					
<i>**Medication orders can be placed with Accredo via E-prescribe - Accredo (1620 Century Center Pkwy, Memphis, TN 38134-8822   NCPDP 4436920), Fax 888.302.1028, or Verbal 866.759.1557</i>					
<b>Facility and/or doctor dispensing and administering medication:</b> Facility Name: State: Tax ID#: Address (City, State, Zip Code): <b>Where will this drug be administered?</b> <input type="checkbox"/> Patient's Home <input type="checkbox"/> Physician's Office <input type="checkbox"/> Hospital Outpatient <input type="checkbox"/> Other (please specify):  <b>NOTE:</b> Per some Cigna plans, infusion of medication MUST occur in the least intensive, medically appropriate setting. Is this patient a candidate for re-direction to an alternate setting (such as alternate infusion site, physician's office, home) with assistance of a Specialty Care Options Case Manager? <input type="checkbox"/> Yes <input type="checkbox"/> No (provide medical necessity rationale):					
Is the requested medication for a chronic or long-term condition for which the prescription medication may be necessary for the life of the patient? <input type="checkbox"/> Yes <input type="checkbox"/> No					
<b>Diagnosis related to use:</b> <input type="checkbox"/> Hereditary angioedema (HAE) due to C1 inhibitor (C1-INH) deficiency <input type="checkbox"/> Hereditary angioedema (HAE) with normal C1 inhibitor (C1-INH) - Please note: This is also known as HAE type III. <input type="checkbox"/> other					

## Clinical Information:

(if HAE due to C1-INH deficiency) What type of hereditary angioedema (HAE) does the patient have?

- ☐ HAE due to C1 esterase inhibitor deficiency, type I  
☐ HAE due to C1 esterase inhibitor deficiency, type II  
☐ Other

(if HAE due to C1-INH deficiency type I or II) What is the indication being requested?

- ☐ Prophylaxis of hereditary angioedema (HAE) attacks  
☐ Treatment of acute hereditary angioedema (HAE) attacks  
☐ Other

(if HAE due to C1-INH deficiency PROPHY) Is this medication being prescribed by, or in consultation with, an allergist/immunologist or a physician who specializes in the treatment of Hereditary Angioedema (HAE) or related disorders?

☐ Yes ☐ No

(if HAE due to C1-INH deficiency PROPHY) Is the patient currently receiving Berinert or Cinryze for Hereditary Angioedema (HAE) prophylaxis?

☐ Yes ☐ No

(if yes) Has the patient had prior approval through the Cigna Coverage Review Department for this product? - Please note: If the patient is currently receiving the requested therapy but has not previously received approval of Berinert or Cinryze for this specific indication through the Cigna Coverage Review Department, review under criteria for Initial Therapy (answer "No" to this question).

☐ Yes ☐ No

(if HAE due to C1-INH deficiency TREATMENT) Is this medication being prescribed by, or in consultation with, an allergist/immunologist or a physician who specializes in the treatment of Hereditary Angioedema (HAE) or related disorders?

☐ Yes ☐ No

(if HAE due to C1-INH deficiency TREATMENT) Had the patient treated previous acute Hereditary Angioedema (HAE) attacks with Berinert or Cinryze?

☐ Yes ☐ No

(if yes) Has the patient had prior approval through the Cigna Coverage Review Department for this product? - Please note: If the patient is currently receiving the requested therapy but has not previously received approval of Berinert or Cinryze for this specific indication through the Cigna Coverage Review Department, review under criteria for Initial Therapy (answer "No" to this question).

☐ Yes ☐ No

(if HAE due to C1-INH deficiency PROPHY - currently receiving with prior approval from Cigna) Is documentation being provided to confirm the patient's Hereditary Angioedema (HAE) (type I or type II) diagnosis? - Please note: a diagnosis of HAE with normal C1-INH (also known as HAE type III) does NOT satisfy this requirement. PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes ☐ No

(if HAE due to C1-INH deficiency PROPHY - currently receiving with prior approval from Cigna) According to the prescriber, has the patient had a favorable clinical response since initiating Berinert or Cinryze compared with baseline (that is, prior to initiating prophylactic therapy)? Please Note: Examples of favorable clinical response include decrease in HAE acute attack frequency, decrease in HAE attack severity, or decrease in duration of HAE attacks.

☐ Yes ☐ No

(if HAE due to C1-INH deficiency TREATMENT - previously treated with prior approval from Cigna) Is documentation being provided to confirm the patient's Hereditary Angioedema (HAE) (type I or type II) diagnosis? - Please note: a diagnosis of HAE with normal C1-INH (also known as HAE type III) does NOT satisfy this requirement. PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes ☐ No

(if HAE due to C1-INH deficiency TREATMENT - previously treated with prior approval from Cigna) According to the prescriber, has the patient had a favorable clinical response with Berinert or Cinryze being used as treatment? Please Note: Examples of favorable clinical response include decrease in the duration of HAE attacks, quick onset of symptom relief, complete resolution of symptoms, or decrease in HAE acute attack frequency or severity.

☐ Yes ☐ No

(if HAE due to C1-INH deficiency PROPHY- not currently receiving OR no prior approval thru Cigna) Is documentation being provided to show that the patient's Hereditary Angioedema (HAE) (type I or type II) has been confirmed by low levels of functional C1-INH protein (less than 50% of normal) at baseline, as defined by the laboratory reference values? PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes ☐ No

(if yes) Is documentation being provided to show that the patient's Hereditary Angioedema (HAE) (type I or type II) has been confirmed by lower than normal serum C4 levels at baseline, as defined by the laboratory reference values? PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information. ☐ Yes ☐ No

(if HAE w/normal C1-INH) What type of hereditary angioedema (HAE) does the patient have?

- ☐ HAE with normal C1 esterase inhibitor, type III  
☐ Other

(if HAE w/normal C1-INH, type III) What is the indication being requested?

- ☐ Treatment of acute hereditary angioedema (HAE), type III attacks  
☐ Prophylaxis of hereditary angioedema (HAE), type III attacks  
☐ Other

(if treatment of acute HAE, type III attacks) Had the patient treated previous acute Hereditary Angioedema (HAE) attacks with Berinert or Cinryze? ☐ Yes ☐ No

(if yes) Has the patient had prior approval through the Cigna Coverage Review Department for this product? - Please note: If the patient is currently receiving the requested therapy but has not previously received approval of Berinert or Cinryze for this specific indication through the Cigna Coverage Review Department, review under criteria for Initial Therapy (answer "No" to this question). ☐ Yes ☐ No

(if HAE w/normal C1-INH - previously treated with prior approval from Cigna) Is documentation being provided to confirm the patient's diagnosis of hereditary angioedema (HAE) with normal C1-INH (type III)? - PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information. ☐ Yes ☐ No

(if HAE w/normal C1-INH - previously treated with prior approval from Cigna) According to the prescriber, has the patient had a favorable clinical response with Berinert or Cinryze treatment? - Please note: Examples of a favorable clinical response include decrease in the duration of HAE attacks, quick onset of symptom relief, complete resolution of symptoms, or decrease in HAE acute attack frequency or severity. ☐ Yes ☐ No

(if HAE w/normal C1-INH - previously treated with prior approval from Cigna) Is this medication being prescribed by, or in consultation with, an allergist/immunologist or a physician who specializes in the treatment of HAE or related disorders? ☐ Yes ☐ No

(if HAE w/normal C1-INH - not previously treated OR no prior approval from Cigna) Is this medication being prescribed by, or in consultation with, an allergist/immunologist or a physician who specializes in the treatment of Hereditary Angioedema (HAE) or related disorders? ☐ Yes ☐ No

(if HAE w/normal C1-INH - not previously treated OR no prior approval from Cigna) According to the prescriber, are the recurrent angioedema attacks responsive to high-dose oral H1 antihistamine therapy? - Please note: High dose oral H1 antihistamine therapy is the highest dose tolerated by the patient and can be up to four times the FDA approved dose. ☐ Yes ☐ No

(if HAE w/normal C1-INH - not previously treated OR no prior approval from Cigna) Is documentation being provided to confirm the patient has normal levels of C1-INH (protein level and/or functional activity), as defined by the laboratory reference values? - PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information. ☐ Yes ☐ No

(if HAE w/normal C1-INH - not previously treated OR no prior approval from Cigna) Is documentation being provided to confirm the patient has normal serum C4 levels, as defined by the laboratory reference values? - PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information. ☐ Yes ☐ No

(if HAE w/normal C1-INH - not previously treated OR no prior approval from Cigna) Is documentation being provided to confirm the patient has a confirmed pathogenic variant in ONE of the following: factor XII (F12), plasminogen (PLG), angiopoietin-1 (ANGPT1), kininogen-1 (KNG1), myoferlin (MYOF), and heparan sulfate glucosamine 3-O-sulfotransferase-6 (HS3OST6)? - PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes ☐ No

(if no) Is documentation being provided to confirm that a pathogenic variant has not been identified? - PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes ☐ No

(if yes) Does the patient have a known family history of hereditary angioedema (HAE) with normal C1 inhibitor?

☐ Yes ☐ No

(if no) Does the patient have a family history of recurrent angioedema without hives?

☐ Yes ☐ No

(if HAE due to C1-INH deficiency TREATMENT - not previously treated OR no prior approval from Cigna) Is documentation being provided to show that the patient's Hereditary Angioedema (HAE) (type I or type II) has been confirmed by low levels of functional C1-INH protein (less than 50% of normal) at baseline, as defined by the laboratory reference values? PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes ☐ No

(if yes) Is documentation being provided to show that the patient's Hereditary Angioedema (HAE) (type I or type II) has been confirmed by lower than normal serum C4 levels at baseline, as defined by the laboratory reference values? PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes ☐ No

(if Berinert and for HAE due to C1-INH deficiency TREATMENT) Does the patient have a history of at least one laryngeal attack, as per the prescriber?

☐ Yes ☐ No

(if no) Does the patient have an allergy to rabbits or rabbit-derived products?

☐ Yes ☐ No

(if no) Is documentation being provided to confirm that the patient has tried Ruconest? PLEASE NOTE: Medical documentation specific to your response must be attached to this case or your request may be denied. Documentation may include, but is not limited to, chart notes, laboratory results, medical test results, claims records, prescription receipts, and/or other information. All documentation must include patient-specific identifying information.

☐ Yes

☐ No - The preferred product is Ruconest. You may review for this product.

#### Additional pertinent information:

Attestation: I attest the information provided is true and accurate to the best of my knowledge. I understand that the Health Plan or insurer its designees may perform a routine audit and request the medical information necessary to verify the accuracy of the information reported on this form.

Prescriber Signature: \_\_\_\_\_ Date: \_\_\_\_\_

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