

Pharmacy Drug Policy Checklist

POLICY NAME Hereditary Angioedema POLICY # 2608P

Criteria

Coverage Criteria for Acute Attacks (Berinert, Firazyr, icatibant, Sajazir, Kalbitor, Ruconest)	
	Diagnosis of C1INH-HAE (formerly types I/II)
	• Evidence of low C4 level (<14 mg/dL AND evidence of low C1 inhibitor (C1INH <19.9 mg/dL)
	OR low C1INH functional level (functional C1INH < 72%)
	Prescribed by a(n) allergist (allergy doctor), immunologist (immune system doctor), or
	rheumatologist (musculoskeletal doctor)
	Appropriate age per approved FDA labeling
	Presence of acute hereditary angioedema (HAE) attack confirmed by one or more symptoms
	(airway swelling, severe abdominal pain, facial swelling, throat swelling, nausea and vomiting, painful face distortion)
	Medications associated with angioedema (e.g., ACE inhibitors, ARBs, NSAIDs, estrogens) have
	been evaluated and, if appropriate, discontinued
	Request for coverage is reviewed by both a pharmacist and a medical director
	Initial Approval: 12 months, Reapproval: 12 months with documented clinical benefit
Coverage Criteria for Prophylaxis (Cinryze, Haegarda, Orladeyo, Takhzyro)	
	Diagnosis of C1INH-HAE (formerly types I/II)
	Appropriate age per approved FDA labeling
	History of at least one HAE attack, without the presence of urticaria (raised, itchy rash), per month
	Prescribed by a(n) allergist (allergy doctor), immunologist (immune system doctor), or rheumatologist (musculoskeletal doctor)
	Medications associated with angioedema (e.g., ACE inhibitors, ARBs, NSAIDs, estrogens) have been evaluated and, if appropriate, discontinued
	Trial and failure, intolerance, or contraindication to any one of the following:

alpha-alkylated androgens (stanozolol, danazol), OR Anti-fibrinolytic agents (aminocaproic acid, tranexamic acid)	
	Request for coverage is reviewed by both a pharmacist and a medical director
	Initial Approval: 12 months, Reapproval: 12 months with documented clinical benefit
Exclusion Criteria – Any of the following prevents coverage	
	Treatment of any other form of chronic, recurrent angioedema, such as: hereditary angioedema with normal C1 inhibitor (formerly Type III HAE), acquired angioedema, idiopathic angioedema, or recurrent angioedema associated with urticaria and all other indications because its effectiveness has not been established
	Medications for acute attacks will not be approved if being used in combination with another medication used for the treatment of acute attacks because this combination lacks strong evidence for use
	Medications for prophylaxis will not be approved if being used in combination with another medication used for the prevention of attacks because this combination lacks strong evidence for use Drug FDA Approved Dose MDL Berinert (human C1 esterase inhibitor) ≥5 y/o