PROVIDER NOTIFICATION OF POLICY CRITERIA CHANGE					
POLICY TITLE	**POLICY NUMBER**	**CRITERIA CHANGE**	**MATERIAL AMENDEMENT**	**EFFECTIVE DATE**	**LINK TO FULL POLICY**
Betibeglogene autotemcel (e.g., Zynteglo)	2022046	Coverage criteria update. 1. Adult or pediatric individual has documented diagnosis of Beta-thalassemia as evidenced by one of the following genotypes confirmed by globin gene testing (Zynteglo, 2022; Locatelli, 2022): a. Beta0/Beta0; **OR** b. Beta0/Beta0 – like (see policy guidelines); **OR** c. Non-Beta0/Beta0 (see policy guidelines); **AND** 2. Individual requires regular peripheral blood transfusions to maintain target hemoglobin levels as defined by documentation of the following: a. History of receiving transfusions of greater than or equal to 100 ml per kilogram of body weight of packed red blood cells per year; **OR** b. History of receiving greater than or equal to 8 transfusions per year in the prior 2 years at the time of treatment decision (Locatelli, 2022); **AND** 3. *Applicable only to individuals less than 18 years of age:* Individual does not have an available and willing human leukocyte antigen-identical or human leukocyte antigen-matched donor. (Locatelli, 2022) (Kassim 2024); **AND** 4. Individual has not received allogenic hematopoietic stem cell transplant (Locatelli, 2022); **AND** 5. Individual meets the institutional requirements for a stem cell transplant procedure where the individual is expected to receive gene therapy (see policy guidelines): a. Adequate Karnofsky performance status or Lansky performance status	No	1/21/2026	https://secure.arkansasbluecross.com/members/report.aspx?policyNumber=2022046

		b. Absence of advanced liver disease c. Adequate estimated glomerular filtration rate (eGFR) d. Adequate diffusing capacity of the lungs for carbon monoxide (DLCO) e. Adequate heart function f. Absence of clinically significant active infection(s) 6. Individual does not have a history of receiving gene therapy or is not under consideration for treatment with another gene therapy for beta thalassemia			
Hereditary Angioedema (HAE) with normal C1 inhibitor levels, Prophylaxis and Acute Treatment	2024073	Coverage criteria updated for C1 Esterase Inhibitor (e.g., Berinert) and C1 Esterase Inhibitor, Recombinant (e.g., Ruconest). C1 Esterase Inhibitor (e.g., Berinert) **INITIAL APPROVAL:** 1. Individual is a child, adolescent or adult with a diagnosis of hereditary angioedema; **AND** 2. Individual has a normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria: a. Individual has an F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation as confirmed by genetic testing; **OR** b. Individual has a documented family history of angioedema, and the angioedema was refractory to a trial of high-dose antihistamine therapy (i.e., cetirizine at 40 mg per day or the equivalent) for at least one month; **AND** 3. Individual has a documented history of recurrent angioedema in the absence of concomitant urticaria with moderate to severe attacks such as airway swelling, severe abdominal pain, facial swelling,	No	1/21/2026	https://secure.arkansasbluecross.com/members/report.aspx?policyNumber=2024073

		nausea and vomiting, or painful facial distortion; **AND** 4. C1 esterase inhibitor (e.g., Berinert) is being used for treatment of acute abdominal, facial, or laryngeal attacks (not prophylaxis). C1 Esterase Inhibitor, Recombinant (e.g., Ruconest) **INITIAL APPROVAL:** 1. Individual is an adolescent or adult with a diagnosis of hereditary angioedema; **AND** 2. Individual has a normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria: a. Individual has an F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation as confirmed by genetic testing; **OR** b. Individual has a documented family history of angioedema, and the angioedema was refractory to a trial of high-dose antihistamine therapy (i.e., cetirizine at 40 mg per day or the equivalent) for at least one month; **AND** 3. Individual has a documented history of recurrent angioedema in the absence of concomitant urticaria with moderate to severe attacks such as airway swelling, severe abdominal pain, facial swelling, nausea and vomiting, or painful facial distortion; **AND** 4. Ruconest is being used for treatment of acute attacks of non-laryngeal origin, (not prophylaxis).			