BOSTON MEDICAL CENTER HEALTHNet PLAN

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Pharmacy Policy

Homozygous Familial Hypercholesterolemia

Policy Number: 9.603
Version Number: 2.0
Version Effective Date: 3/1/2022

Product Applicability	☐ **All Plan+ Products**
Well Sense Health Plan ☐ New Hampshire Medicaid	**Boston Medical Center HealthNet Plan** ☐ MassHealth - MCO ☐ MassHealth - ACO ☒ Qualified Health Plans/ConnectorCare/Employer Choice Direct ☐ Senior Care Options

Note: Disclaimer and audit information is located at the end of this document.

Prior Authorization Policy

Products Affected:

- **Evkeeza (evinacumab-dgnb)**
- **Juxtapid (lomitapide)**

The Plan may authorize coverage of the above products for members meeting the following criteria:

Covered Use	All FDA approved indications not otherwise excluded
Exclusion Criteria	Concurrent use with PCSK9 inhibitors
Required Medical Information	**Juxtapid:** 1. Documentation of diagnosis of homozygous familial hypercholesterolemia; confirmed by the following (A AND B): a.

+ *Plan* refers to Boston Medical Center Health Plan, Inc. and its affiliates and subsidiaries offering health coverage plans to enrolled members. The Plan operates in Massachusetts under the trade name Boston Medical Center HealthNet Plan and in other states under the trade name Well Sense Health Plan.

i. Genetic testing confirming mutation of two mutant alleles at low-density lipoprotein receptor (LDLR), apolipoprotein B (APOB), proproteinconvertase subtilisinkexintype 9 (PCSK9), or low-density lipoprotein receptor adaptor protein 1 (LDLRAP1) gene locus; OR

ii. If genetic testing was negative or inconclusive: prescriber must provide documentation attesting to a mutation that may cause familial hypercholesterolemia

b. One of the recognized diagnostic criteria are met

i. Dutch Lipid Clinic criteria; OR

ii. Simon Broome criteria

2. Documentation of medication being prescribed by, or in written consultation with (documentation required), a lipid specialist (e.g., cardiologist, endocrinologist, lipid specialist); AND
3. **One** of the following:
    a. Inadequate LDL reduction while adherent to a minimum of 90 day continuous use of atorvastatin 80mg or rosuvastatin 40mg in combination with ezetimibe evidenced by:
        i. Current LDL-C greater than or equal to 100mg/dL; ***AND***
        ii. Less than a 50 percent reduction in LDL-C from baseline; ***OR***
    b. Inability to tolerate a high intensity statin (atorvastatin 80mg or rosuvastatin 40mg); ***AND***
        i. Inadequate response while adherent to a minimum of 90 day continuous use of a maximum tolerated dose of a non-high intensity statin and ezetimibe evidenced by:
            1. Current LDL-C greater than or equal to 100mg/dL; ***AND***
4. An inadequate response to Repatha while adherent to a minimum of 90 day continuous use as evidenced by:
    a. Current LDL-C greater than or equal to 100 mg/dL; ***OR***
    b. An adverse effect or contraindication to Repatha; ***AND***
5. Baseline LDL-C level is provided

Evkeeza:

1. Documentation of medication being prescribed by, or in written consultation with (documentation required), a lipid specialist (e.g., cardiologist, endocrinologist, lipid specialist); AND
2. Member is greater than or equal to 12 years of age; AND
3. Baseline LDL-C level is provided; AND
4. Documentation of confirmed diagnosis of homozygous hypercholesterolemia confirmed by the following (A AND B):
    a.

	i. Genetic testing confirming mutation of two mutant alleles at low-density lipoprotein receptor (LDLR), apolipoprotein B (APOB), proproteinconvertase subtilisinkexintype 9 (PCSK9), or low-density lipoprotein receptor adaptor protein 1 (LDLRAP1) gene locus; OR ii. If genetic testing was negative or inconclusive: prescriber must provide documentation attesting to a mutation that may cause familial hypercholesterolemia b. One of the recognized diagnostic criteria are met i. Dutch Lipid Clinic criteria; OR ii. Simon Broome criteria 5. Documentation of inadequate response to or contraindication to ALL of the following despite 90 days of compliant therapy: a. One high-intensity statin prescribed at the maximally tolerate dose; AND b. Ezetimibe; AND c. PSCK9 inhibitor 6. Attestation that current lipid lowering therapy will be used concurrently; AND 7. Attestation of patient lifestyle modifications (e.g. low fat diet, exercise)
Age Restriction	Juxtapid: 18 years of age and older Evkeeza: 12 years of age and older
Prescriber Restriction	Prescribed by, or in written consultation with (documentation required), a lipid specialist (e.g., cardiologist, endocrinologist, lipid specialist
Coverage Duration	Initial: 6 months Reauthorization: 12 months
Other criteria	Re-authorization: 1. Initial criteria has been met; ***AND*** 2. Documentation of clinical response to therapy as defined by a decrease in LDL-C levels from baseline; ***AND*** 3. Member has been compliant with therapy (confirmed by pharmacy claims); ***AND*** 4. For Juxtapid: Provider attests that liver function test including AST and ALT are being monitored every 3 months and will be discontinued if liver function tests are 3 times the ULN.

Clinical Background Information and References

1. Rosenson R, de Ferranti S, Durrington P. Treatment of drug resistant hypercholesterolemia. UptoDate. Last updated January 30, 2013, Accessed April 2014. Available from http://www.uptodate.com.
2. Rosenson R, de Ferranti S, Durrington P. Inherited disorders of LDL-cholesterol metabolism. UptoDate. Last updated February 12, 2014, Accessed April 2014. Available from http://www.uptodate.com.
3. Ito M, McGowan M, Moriarty P. Management of familial hypercholesterolemias in adult patients: recommendations from the National Lipid Association Expert Panel on Familial Hypercholesterolemia. Journal of Clinical Lipidology. 2011;5: S38-S45.

4. Juxtapid™ [package insert]. Cambridge (MA): Aegerion Pharmaceuticals; March 2016.
5. Evkeeza [package insert]. Tarrytown, NY; Regeneron Pharmaceuticals; February 2021.

Applicable Coding:

J -Code	Medication
J1305	Injection, evinacumab-dgnb, 5 mg

Original Approval Date	Original Effective Date	Policy Owner	Approved by
12/1/2020	1/1/2021	Pharmacy Services	Pharmacy & Therapeutics (P&T) Committee

Policy Revisions History			
Review Date	Summary of Revisions	Revision Effective Date	Approved by
12/1/2020	Dicontinued policy 9.039 and created new policy for QHP.Kynamro removed from policy and moved to NP for QHP.	1/1/2021	P&T Committee
11/11/2021	Diagnosis documentation expanded to require genetic testing and diagnostic criteria. Baseline LDL-C level is now required to be provided for initial approval. Age restrictions updated to labeling, provider restriction added. Initial approval set to 6 months. Reauthorization criteria now requires initial criteria to have been met. Criteria for Evkeeza added to policy along with Jcode.	3/1/2022	P&T Committee

Next Review Date

11/2022

Other Applicable Policies

Reference to Applicable Laws and Regulations, If Any

Disclaimer Information

Medical Policies are the Plan's guidelines for determining the medical necessity of certain services or supplies for purposes of determining coverage. These Policies may also describe when a service or supply is considered experimental or investigational, or cosmetic. In making coverage decisions, the Plan uses these guidelines and other Plan Policies, as well as the Member's benefit document, and when appropriate, coordinates with the Member's health care Providers to consider the individual Member's health care needs.

Plan Policies are developed in accordance with applicable state and federal laws and regulations, and accrediting organization standards (including NCQA). Medical Policies are also developed, as appropriate, with consideration of the medical necessity definitions in various Plan products, review of current literature, consultation with practicing Providers in the Plan's service area who are medical experts in the particular field, and adherence to FDA and other government agency policies. Applicable state or federal mandates, as well as the Member's benefit document, take precedence over these guidelines. Policies are reviewed and updated on an annual basis, or more frequently as needed. Treating providers are solely responsible for the medical advice and treatment of Members.

The use of this Policy is neither a guarantee of payment nor a final prediction of how a specific claim(s) will be adjudicated. Reimbursement is based on many factors, including member eligibility and benefits on the date of service; medical necessity; utilization management guidelines (when applicable); coordination of benefits; adherence with applicable Plan policies and procedures; clinical coding criteria; claim editing logic; and the applicable Plan – Provider agreement.