SmartPA Criteria Proposal

Drug/Drug Class:	Homozygous Familial Hypercholesterolemia (HoFH) Agents PDL Edit
First Implementation Date:	January 29, 2014
Proposal Date:	May 11, 2021
Prepared For:	MO HealthNet
Prepared By:	MO HealthNet/Conduent
Criteria Status:	☐Existing Criteria ☒Revision of Existing Criteria ☐New Criteria

Executive Summary

Purpose: Ensure appropriate utilization and control of agents for Homozygous Familial Hypercholesterolemia (HoFH)

Why Issue Selected: Familial Hypercholesterolemia (FH) is a genetic disorder characterized by high cholesterol levels, specifically very high levels of low-density lipoprotein (LDL) in the blood. Patients who have one abnormal copy of the LDLR gene have the heterozygous form while those patients who have two abnormal copies of the LDLR gene have the homozygous form. Heterozygous FH is a common genetic disorder occurring in 1:500 people while Homozygous FH (HoFH) is much rarer, occurring in 1 in a million births. Patients with HoFH have severely elevated levels of LDL-C. Physical findings of HoFH may include premature coronary artery disease (CAD) and tendon and skin xanthomas. Treatment involves early and aggressive lipid-lowering therapies and lipoprotein apheresis. Patients with HoFH are typically less responsive to standard lipid-lowering therapies including high-intensity statins and proprotein convertase subtilisin/kexin type 9 (PCSK9) inhibitors. Some patients with HoFH are non-responders to standard therapy.

Juxtapid® is indicated as an adjunct to lipid-lowering medications, treatments, and diet to reduce low-density lipoprotein-cholesterol (LDL-C), apolipoprotein B, total cholesterol (TC) and non-high density lipoprotein-cholesterol (non HDL-C) in patients with homozygous familial hypercholesterolemia (HoFH).

Evkeeva™ is an angiopoietin-like 3 (ANGPTL3) inhibitor indicated as an adjunct to other LDL-C lowering therapies for the treatment of adult and pediatric patients, aged 12 years and older, with HoFH.

Total program savings for the PDL classes will be regularly reviewed.

Program-Specific Information:

Date Range FFS 1-01-2020 to 12-31-2020			
Drug	**Claims**	**Cost per unit (MAC)**	**Cost per year (MAC)**
JUXTAPID 5 MG CAPSULE	0	$1,590.52 per capsule	$534,414.72
JUXTAPID 10 MG CAPSULE	0		
JUXTAPID 20 MG CAPSULE	0		
JUXTAPID 30 MG CAPSULE	0		
JUXTAPID 40 MG CAPSULE	0		
JUXTAPID 60 MG CAPSULE	0		
EVKEEZA 345 MG/2.3 ML VIAL	0	$4,668.75 per ml	$448,200.00 (based on an 80 kg pt)
EVKEEZA 1,200 MG/8 ML VIAL	0		

Type of Criteria: ☐ **Increased risk of ADE** ☒ **Preferred Drug List** ☒ **Appropriate Indications** ☐ **Clinical Edit**

Data Sources: ☐ **Only Administrative Databases** ☒ **Databases + Prescriber-Supplied**

Setting & Population

- Drug class for review: Homozygous Familial Hypercholesterolemia (HoFH) Agents
- Age range: All appropriate MO HealthNet participants aged **12** years or older

Approval Criteria

- **Prescribed by or in consultation with a lipid disorder specialist or other appropriate specialist in the treated disease state AND**
- Participant is currently not pregnant **AND**
- Documented diagnosis of Homozygous Familial Hypercholesterolemia **confirmed by genetic testing AND**
- Documentation of LDL-C > 175 mg/dl **AND**
- Documented therapeutic trial of a high intensity statin or documented ADE/ADR to high intensity statin therapy **AND**
- **Documented therapeutic trial of a PCSK9 inhibitor (Repatha or Praluent) AND**
- For Juxtapid: Participant is aged 18 years or older
- **For Evkeeza: Participant is aged 12 years or older**

Denial Criteria

- Therapy will be denied if all approval criteria are not met
- **For Juxtapid:**
  - Documented diagnosis of moderate or severe hepatic impairment
  - Dose on claim exceeds 60 mg per day

Required Documentation

Laboratory Results:	X	Progress Notes:	
MedWatch Form:		Other:	

Disposition of Edit

Denial: Exception Code “0682” (Clinical Edit)
Rule Type: CE

Default Approval Period

1 year

References

1. Evidence-Based Medicine and Fiscal Analysis: “Homozygous Familial Hypercholesterolemia Products – Therapeutic Class Review”, Conduent Business Services, L.L.C., Richmond, VA; July 2020.
2. Evidence-Based Medicine Analysis: “Homozygous Familial Hypercholesterolemia Products”, UMKC-DIC; July 2020.
3. Juxtapid (lomitapide) [package insert]. Dublin, Ireland: Amryt Pharmaceuticals DAC; September 2020.
4. Evkeeza (evinacumab-dgnb) [package insert]. Tarrytown, NY: Regeneron Pharmaceuticals; 2021.
5. Lippincott, Williams, Wilkins. PDR Electronic Library, Montvale NJ; 2021.
6. USPDI, Micromedex; 2021.
7. Facts and Comparisons eAnswers (online); 2021 Clinical Drug Information, LLC.