SmartPA Criteria Proposal

Drug/Drug Class:	Homozygous Familial Hypercholesterolemia (HoFH) Agents PDL Edit
First Implementation Date:	January 29, 2014
Proposed Date:	December 16, 2021
Prepared For:	MO HealthNet
Prepared By:	MO HealthNet/Conduent
Criteria Status:	☐Existing Criteria ☒Revision of Existing Criteria ☐New Criteria

Executive Summary

Purpose: The MO HealthNet Pharmacy Program will implement a state-specific preferred drug list.

Why Issue Selected: Familial Hypercholesterolemia (FH) is a genetic disorder characterized by high cholesterol levels, specifically high levels of low-density lipoprotein-cholesterol (LDL-C) in the blood. Patients who have one abnormal copy of the low-density lipoprotein receptor (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 a branded drug product indicated as an adjunct to lipid-lowering medications, treatments, and diet to reduce LDL-C, apolipoprotein B, total cholesterol (TC) and non-high density lipoprotein-cholesterol (non HDL-C) in patients with HoFH.

Evkeeva® is an angiopoietin-like 3 (ANGPTL3) inhibitor indicated as 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:

Preferred Agents	Non-Preferred Agents
	• Evkeeza® • Juxtapid®

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

Approval Criteria

- Documented diagnosis of homozygous familial hypercholesterolemia **confirmed by:**
  - **Genetic testing OR**
  - **American Heart Association clinical criteria 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 compliance to the following therapies:**
  - **PCSK9 inhibitor therapy (defined as 90/120 days) AND**
  - **High intensity statin therapy (defined as 90/120 day) OR**
  - **Documented ADE/ADR to high intensity statin therapy AND**
- **Documentation of LDL-C lab result not meeting goal while on therapy with high-intensity statin and PCSK9 inhibitor**
- **For Evkeeza: Participant aged 12 years or older**
- For Juxtapid: Participant aged 18 years or older

Denial Criteria

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

Required Documentation

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

Disposition of Edit

Denial: **Exception Code "0160" (Preferred Drug List)**
Rule Type: **PDL**

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 2021.
2. Evidence-Based Medicine Analysis: “Homozygous Familial Hypercholesterolemia Products”, UMKC-DIC; July 2021.
3. Evkeeza (evinacumab-dgnb) [package insert]. Tarrytown, NY: Regeneron Pharmaceuticals; February 2021
4. Juxtapid (lomitapide) [package insert]. Dublin, Ireland: Amryt Pharmaceuticals DAC; September 2020.
5. Facts and Comparisons eAnswers (online); 2021 Clinical Drug Information, LLC.