Custom Criteria for
BlueCross Blue Shield of Rhode Island Commercial

JUXTAPID/KYNAMRO

GPI CODING:

394800502001**
395000401020**

DESCRIPTION:

Lomitapide

- Indicated as an adjunct to a low-fat diet and other lipid-lowering treatments, including LDL apheresis where available, to reduce low-density lipoprotein cholesterol (LDL-C), total cholesterol (TC), apolipoprotein B (apo B), and non-high-density lipoprotein cholesterol (non-HDL-C) in patients with HoFH.
- The FDA granted Juxtapid orphan drug status for familial chylomicronemia and HoFH.
- Limitations of Use: the safety and effectiveness of lomitapide have not been established in patients with hypercholesterolemia who do not have HoFH and the effect of lomitapide on cardiovascular morbidity and mortality has not been determined.

Mipomersen

- Indicated as an adjunct to lipid-lowering medications and diet to reduce low density lipoprotein-cholesterol (LDL-C), apolipoprotein B (apo B), total cholesterol (TC), and non-high density lipoprotein-cholesterol (non HDL-C) in patients with HoFH.
- Limitations of Use: the safety and effectiveness of KYNAMRO have not been established in patients with hypercholesterolemia who do not have HoFH, the effect of KYNAMRO on cardiovascular morbidity and mortality has not been determined, and the use of KYNAMRO as an adjunct to LDL apheresis is not recommended.

APPROVAL DURATION:

Approval duration: 1 year

Kynamro quantity limit: 1 vial/week

Juxtapid quantity limits:
- 5 mg tablets: 1 tablet/day
- 10 mg tablets: 1 tablet/day
- 20 mg tablets: 3 tablets/day

CRITERIA FOR JUXTAPID & KYNAMRO

Patient has a diagnosis of homozygous familial hypercholesterolemia (HoFH) as evidence by

1. Genetic confirmation of 2 mutant alleles at the LDL receptor, ApoB, PCSK9, or autosomal recessive hypercholesterolemia (ARH) adaptor protein gene locus

OR

2. Untreated LDL >650 mg/dL or treated LDL >300 mg/dL or treated non-HDL >330 mg/dL with at least one of the following:
   a. Cutaneous or tendonous xanthoma before age 10 years

b. History of early vascular disease (men <55 years of age, women <60 years of age) on both sides of the family if parental LDL levels are unknown
c. Elevated LDL cholesterol levels before lipid-lowering therapy consistent with heterozygous FH in both parents where LDL levels are known:
    a. LDL cholesterol >250 mg/dL in a patient aged 30 or more;
    b. LDL cholesterol >220 mg/dL for patients aged 20 to 29;
    c. LDL cholesterol >190 mg/dL in patients under age 20;

AND
Juxtapid or Kynamro will be used as adjunct to a low-fat diet and other lipid-lowering treatments AND
Patient does not have any of the following contraindications to therapy:

a. Pregnancy (Juxtapid only)
b. Concomitant use with strong or moderate CYP3A4 inhibitors (Juxtapid only)
c. Moderate or severe hepatic impairment or active liver disease including unexplained persistent abnormal liver function tests

AND
Patient has tried and had an inadequate response after compliance to maximally tolerated doses for a minimum of 8 weeks or contraindication to TWO statins AND
Patient has tried and had an inadequate response after compliance for a minimum of 8 weeks to at least one of the following: ezetimibe, cholestyramine, OR lipid apheresis

Authorization for continued use shall be reviewed at least every 12 months to confirm the following: patient does not have any contraindications to therapy and has had at least a 10% reduction in LDL while on therapy.