Utilization Management Policy Name: Juxtapid - Kynamro

Restricted Product(s):
- JUXTAPID (lomitapide mesylate)
- KYNAMRO (mipomersen sodium)

FDA Approved Use:
- Juxtapid
  - 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 homozygous familial hypercholesterolemia (HoFH)
  - The safety and effectiveness of Juxtapid have not been established in patients with hypercholesterolemia who do not have HoFH.
  - The effect of Juxtapid on cardiovascular morbidity and mortality has not been determined
- Kynamro
  - 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 homozygous familial hypercholesterolemia (HoFH)
  - 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.
  - The use of Kynamro as an adjunct to LDL apheresis is not recommended.

Rationale:
These products can be used in attempts to treat conditions that have not been validated by the FDA. This program ensures that members are receiving this medication for conditions that have the appropriate evidence to support its use. Quantity limits have been added to ensure safe and effective use.

Criteria Summary:
FDA approved use/medical necessity; exception to quantity limitation

Criteria for Approval of Restricted Product(s):
Initial Coverage Criteria
1. The patient has a confirmed diagnosis of homozygous familial hypercholesterolemia (HoFH) through ONE of the following [medical documentation is required]:
   a. Genetic confirmation of two mutant alleles at the LDLR, Apo-B, PCSK9, or ARH adaptor protein 1/LDLRAP1 gene locus; OR
   b. Untreated LDL-C >500 mg/dL or treated LDL-C ≥300 mg/dL with one of the following:
      i. Cutaneous or tendon xanthoma before age 10 years; OR
      ii. Untreated elevated LDL-C levels consistent with heterozygous FH in both parents [untreated total cholesterol >290 mg/dL or untreated LDL-C >190 mg/dL]; AND

2. One of the following:
   a. The patient is currently taking and adherent* to high-intensity statin** therapy, OR
   b. The patient is intolerant*** to at least 2 different statins; AND

3. The patient is on one additional lipid-lowering medication (e.g., ezetimibe, nicotinic acid, bile acid sequestrant, or fibrate); AND

4. The patient will be maintained on a low fat diet with <20% of calories from fat; AND

5. The patient will NOT be taking a PCSK9 inhibitor (i.e. Repatha or Praluent) while on Juxtapid or Kynamro therapy; AND

6. If the request is for Juxtapid (lomitapide), the patient is receiving dietary supplement(s) containing fat soluble vitamins (recommended approximately 400IU vitamin E, 210 mg alpha-linolenic acid (ALA), 200 mg linoleic acid, 110 mg eicosapentaenoic acid (EPA), and 80mg docosahexaenoic acid (DHA) per day); AND

7. If the request is for Kynamro ( mipomersen), the patient will not be receiving apheresis while on therapy with Kynamro; AND

8. For formularies that exclude (non-formulary) the requested medication, Non-formulary Exception Criteria applies (outlined below)****

   *Adherence is defined as the proportion of days covered (PDC) to be 80% or greater over the last 6 months
   **High-intensity statin is the equivalent of rosuvastatin 20-40mg or atorvastatin 40-80mg
   ***Intolerance is defined as (1) the inability to tolerate any dose or (2) the inability to increase the dose above the lowest FDA-approved tablet strength

**Duration of Approval:**
   Juxtapid: 365 days
   Kynamro: 24 weeks

Continuation Coverage Criteria

1. The patient has a prior approval for this medication from Blue Cross NC; AND
2. The patient has shown at least a 10% reduction from baseline in at least ONE of the following metrics: LDL-C, Apo B, total cholesterol, Non-HDL-C, Triglycerides; AND

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3. The patient is on a concurrent lipid lowering regimen (e.g. statins, ezetimibe, nicotinic acid, bile acid sequestrant, or fibrate); **AND**
4. The patient will be maintained on a low fat diet with <20% of calories from fat; **AND**
5. If the request is for Juxtapid, the patient is receiving dietary supplement(s) containing fat soluble vitamins (recommended approximately 400IU vitamin E, 210 mg alpha-linolenic acid (ALA), 200 mg linoleic acid, 110 mg eicosapentaenoic acid (EPA), and 80mg docosahexaenoic acid (DHA) per day); **AND**
6. If the request is for Kynamro, the patient will not be receiving apheresis while on therapy with Kynamro.

**Duration of Approval:** 365 days

**Quantity Limitations:** quantity limitations apply to brand and associated generic products.

<table>
<thead>
<tr>
<th>Medication</th>
<th>Quantity per Day (unless specified)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Juxtapid 5mg capsules</td>
<td>2</td>
</tr>
<tr>
<td>Juxtapid 10mg capsules</td>
<td>2</td>
</tr>
<tr>
<td>Juxtapid 20mg capsules</td>
<td>3</td>
</tr>
<tr>
<td>Juxtapid 30mg capsules</td>
<td>2</td>
</tr>
<tr>
<td>Juxtapid 40mg capsules</td>
<td>1</td>
</tr>
<tr>
<td>Juxtapid 60mg capsules</td>
<td>1</td>
</tr>
<tr>
<td>Kynamro 200mg/mL injection</td>
<td>1 injection per week</td>
</tr>
</tbody>
</table>

**Quantity Limit Exception Criteria:**

1. The quantity (dose) requested is for documented titration purposes at the initiation of therapy (authorization for a 90 day titration period); **AND**
2. The prescribed dose cannot be achieved using a lesser quantity of a higher strength; **AND**
3. The quantity (dose) requested does not exceed the maximum FDA labeled dose, when specified, or to the safest studied dose per the manufacturer’s product insert; **OR**
4. If the quantity (dose) requested exceeds the maximum FDA labeled dose, when specified, or to the safest studied dose per the manufacturer’s product insert, then the prescriber must submit documentation in support of therapy with a higher dose for the intended diagnosis (submitted documentation may include medical records OR fax form which reflects medical record documentation that shows the length of time the requested dose has been used, and what other medications and doses have been tried and failed).

**Duration of Approval:** 365 days
**Non-formulary Exception Criteria**

Non-Formulary Exception criteria applies on formularies which exclude requested product(s). Satisfactory completion of criteria points (above) may satisfy some, or all, portions of the Non-Formulary Exception Criteria. This criteria is summarized as:

a) Request must be for an FDA approved indication; **AND**

b) Patient must have a trial and failure of up to **TWO** formulary medications or a clinical contraindication/intolerance to those medications not tried.

**References:** all information referenced is from FDA package insert unless otherwise noted below.

**Policy Implementation/Update Information:** Originated: January 2014; last updated: August 2017.

- Aug 2017: Criteria added in regard to concomitant use of PCSK9 products; removal of additional requirements for Basic Open formulary; removal of contraindication language
- Jun 2015: New to market Juxtapid added to policy
- Jan 2014: Original utilization management criteria issued

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