Pharmacy Drug Policy Checklist

POLICY NAME	Juxtapid (lomitapide)	POLICY #	2389P

Criteria

Initial Coverage for the Treatment of Homozygous Familial Hypercholesterolemia (HoFH)

- ☐ **1.1** Documented diagnosis of homozygous familial hypercholesterolemia, confirmed by one of the following:
  - Documentation of two identified mutations in any one of the following genes: LDL receptor (LDLR), Apo B, or Proprotein convertase subtilisin/kexin type 9 (PCSK9) gene, OR
  - A formal clinical diagnosis of familial hypercholesterolemia made using one of the following tools: ? U.S. Make Early Diagnosis Prevent Early Death (MEDPED) ? Dutch Lipid Clinical Network ? Simon-Broome Registry
- ☐ **1.2** Ordered by a cardiologist (heart doctor) or lipid specialist
- ☐ **1.3** Age 18 or older
- ☐ **1.4** Triglyceride level < 400mg/dL
- ☐ **1.5** Documentation of ACC/AHA 10-year risk calculation of 7.5% or greater
- ☐ **1.6** Lab value of treated low-density lipoprotein (LDL) level greater than 100mg/dL within the last 30 days,
- ☐ **1.7** Documentation to support one of the following:
  - Documented failure on high-intensity statin therapy (atorvastatin 80mg, Crestor 20mg, Crestor 40mg), in combination with ezetimibe, defined as being unable to decrease LDL level by 50% ? Failure is defined as an inability to decrease LDL level by 50%, and corresponding claims history supporting that the member has filled at least 150 days of both medications in the last six (6) months
  - Documented intolerance to high-intensity statin therapy (defined as severe myalgias/muscle aches and/or creatinine kinase levels greater than 10 times the upper limit of the lab reference range), AND documented failure on medium-intensity statin therapy (atorvastatin 10mg, Crestor 5mg, simvastatin

to 40mg, pravastatin 40mg, lovastatin 40mg, fluvastatin XL 80mg, fluvastatin 40mg twice daily, or Livalo 2 to 4mg) used in combination with ezetimibe ? Failure is defined as an inability to decrease LDL level by 50%, and corresponding claims history supporting that the member has filled at least 150 days of both medications in the last six (6) months

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1.8 Documentation to support one of the following:

- Documented failure after 3 doses of Repatha 420mg in combination with high-intensity statin therapy and ezetimibe unless use of these agents is contraindicated, defined as being unable to decrease LDL level by 30% ? Failure is defined as an inability to decrease LDL level by 30%, and corresponding claims history supporting that the member has filled at least 84 days of Repatha in at least four (4) Criteria Statement of the Policy References months
- Member with a confirmed diagnosis of LDL receptor-negative homozygous familial hyperlipidemia will bypass step through Repatha, OR ? Repatha was not effective at lowering LDL levels in these patients per TESLA Part B trial

☐ **1.9** Documentation that the member will discontinue PCSK9 inhibitor therapy prior to the initiation of Juxtapid

- Safety and efficacy of this combination has not been studied

☐ **1.10** Negative urine pregnancy test in female patient of reproductive potential prior to treatment

☐ **1.11** Documentation that liver function tests will be collected every 4 weeks and after each dose escalation during the first year

☐ **1.12** Documentation that liver function test will be collected at least every 3 months and prior to dosage increases after the first year

☐ **1.13** Confirmation that the member will use daily supplements containing vitamin E 400 units, linoleic acid 200mg, alpha-linolenic acid (ALA) 210mg, eicosapentaenoic acid (EPA) 110mg, and docosahexaenoic acid (DHA) 80mg

☐ **1.14** Confirmation that the member will initiate and maintain a low-fat diet supplying < 20% of energy from fat