AvMed

PHARMACY PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-305-671-0200**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process can be delayed.**

Drug Requested: Juxtapid® (lomitapide)

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member AvMed #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Name/Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight (if applicable): _____ **Date weight obtained:** _____

Requested Medication is ONE of the following:

- ❑ New Start
- ❑ Continuation of therapy – provide therapy start date: _____

Recommended Dosage:

- **Adults –** Oral: Initial: 5 mg once daily; after ≥2 weeks of therapy, may increase to 10 mg once daily, as tolerated; then at ≥4-week intervals, may increase to 20 mg once daily, then to 40 mg once daily, and finally to a maximum dose of 60 mg/day as tolerated.
- **Children ≥ 2 years and Adolescents < 18 years –** Oral: Initial dose and titration schedule varies based on age; titrate dose as needed based on patient tolerability, desired low-density lipoprotein cholesterol (LDL-C) levels, and adverse effects. Adjust dose as needed when patient crosses into next age group.

(Continued on next page)

Age Groups	Weeks 0 to 4	Weeks 5 to 8	Weeks 9 to 12	Weeks 13 to 16	Weeks 17 and beyond
2 to < 11 years	2 mg once daily	Continue 2 mg once daily	Increase to 5 mg once daily	Increase to 10 mg once daily	Increase to 20 mg once daily. Max daily dose: 20 mg/day
11 to < 16 years	2 mg once daily	Increase to 5 mg once daily	Increase to 10 mg once daily	Increase to 20 mg once daily	Increase to 40 mg once daily. Max daily dose: 40 mg/day
16 to < 18 years	5 mg once daily	Increase to 10 mg once daily	Increase to 20 mg once daily	Increase to 40 mg once daily	Continue 40 mg once daily. Max daily dose: 40 mg/day

Quantity Limits:

- 2, 5 & 10 mg capsules – 1 capsule per day
- 20 & 30 mg capsules – 2 capsules per day

CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Authorization: 6 months

- ❑ Member must be 2 years of age or older
- ❑ Requested medication is prescribed by or in consultation with a specialist in cardiology, lipidology, or endocrinology
- ❑ Baseline low-density lipoprotein cholesterol (LDL-C), total cholesterol (TC), apolipoprotein B (apo B), and non-high density lipoprotein cholesterol (non-HDL-C) labs must be obtained prior to initiating treatment **(please submit labs from the past 30 days with request)**
- ❑ Member does **NOT** have heterozygous familial hypercholesterolemia (HeFH)
- ❑ Member has a diagnosis of Homozygous Familial Hypercholesterolemia (HoFH) confirmed by at least **ONE** of the following:
  - ❑ Documented DNA test for functional mutation(s) in LDL receptor alleles or alleles known to affect LDL receptor functionality **(submit test results with request)**
  - ❑ Untreated LDL-C > 500 mg/dL **OR** treated LDL-C ≥ 300 mg/dL along with **ONE** of the following
    - ❑ Cutaneous or tendon xanthoma before 10 years of age
    - ❑ Untreated LDL cholesterol levels consistent with heterozygous FH in both parents (> 190 mg/dL)
- ❑ Member has been receiving stable background lipid lowering therapy for at least 4 weeks **(verified by pharmacy paid claims)**

(Continued on next page)

- ❑ Requested therapy will be used in conjunction with a low-fat or heart-healthy diet and other LDL-lowering therapies (e.g., statins, ezetimibe, PCSK9 inhibitors, LDL apheresis)
- ❑ Member has had an unsuccessful 3-month trial of **ALL** the following, unless not FDA-approved for use in the member's age **(verified by pharmacy and medical paid claims and/or submitted genetic test results)**:
  - ❑ Highest available (or maximally tolerated) dose of atorvastatin **OR** rosuvastatin
  - ❑ ezetimibe (Zetia®)
  - ❑ PCSK9 inhibitor indicated for HoFH (e.g., Repatha), unless contraindicated or if pathogenic/likely pathogenic null-null variant (e.g. LDLR, APOB, PCSK9, or LDLRAP1) is confirmed by genetic testing
  - ❑ Evkeeza® (evinacumab)
- ❑ Despite pharmacological treatment with Evkeeza, a PCSK9 inhibitor, maximally tolerated statin therapy, and ezetimibe, the member's LDL cholesterol remains ≥ 100 mg/dL (or ≥ 70 mg/dL for members with clinical atherosclerotic cardiovascular disease [ASCVD]) while on therapy **(submit pre- and post-treatment cholesterol labs)**
- ❑ If there has been **no response** to a PCSK9 inhibitor (e.g. after an adherent trial, labs show LDL-C has remained the same or increased), PCSK9 inhibitor therapy will be discontinued (**NOTE**: PCSK9 inhibitor therapy may be continued with Juxtapid if LDL-C lowering has been demonstrated while on a PCSK9 inhibitor)

***Provider please note: If the member is not able to use the maximum dose of atorvastatin or rosuvastatin due to muscle symptoms, documentation of a causal relationship must be established between statin use and muscle symptoms**

- ❑ Member must meet **ONE** of the following:
  - ❑ Clinical documentation must demonstrate that the member experienced pain, tenderness, stiffness, cramping, weakness, and/or fatigue in addition to **ALL** the following:
    - ❑ Muscle symptoms resolve after discontinuation of statin
    - ❑ Muscle symptoms occurred when re-challenged at a lower dose of the same statin
    - ❑ Muscle symptoms occurred after switching to an alternative statin
    - ❑ Documentation ruling out non-statin causes of muscle symptoms (e.g., hypothyroidism, reduced renal function, reduced hepatic function, rheumatologic disorders, such as polymyalgia rheumatica, steroid myopathy, vitamin D deficiency, or primary muscle disease)
  - ❑ Member has been diagnosed with rhabdomyolysis associated with statin use and the diagnosis is supported by acute neuromuscular illness or dark urine **AND** an acute elevation in creatine kinase (usually > 5,000 IU/L or 5 times the upper limit of normal [ULN])
- ❑ Prescriber and member are enrolled in the Juxtapid REMS program, and provider will monitor the member's liver function during treatment with Juxtapid
- ❑ Member has no contraindications to therapy with Juxtapid (e.g., pregnancy, coadministration with moderate or strong CYP3A4 inhibitors, moderate to severe hepatic impairment, or active liver disease)

(Continued on next page)

- ❑ Prescriber attests member has been counseled to reduce risk of fat-soluble nutrient deficiency by taking daily supplements that contain amounts of vitamin E, alpha-linolenic acid, eicosapentaenoic acid (EHA), and docosahexaenoic acid (DHA) as recommended in the Juxtapid prescribing information
- ❑ The member's dose will **NOT** exceed the age-specific maximum recommended dose

Reauthorization: 12 months. Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

- ❑ Requested therapy continues to be used in conjunction with a low-fat or heart-healthy diet and other LDL-lowering therapies (e.g., statins, ezetimibe, PCSK9 inhibitors, LDL apheresis)
- ❑ Member has experienced a reduction in LDL-C when compared to the initial baseline labs **(submit labs collected within the last 3 months)**
- ❑ Member has **NOT** experienced any unacceptable toxicity from therapy (e.g., elevated transaminases, hepatic steatosis, hepatotoxicity, or intolerable gastrointestinal side effects)
- ❑ Prescriber will continue to monitor the member's liver function during treatment with Juxtapid
- ❑ Member has no contraindications to therapy with Juxtapid (e.g., pregnancy, coadministration with moderate or strong CYP3A4 inhibitors, moderate to severe hepatic impairment, or active liver disease)
- ❑ Prescriber attests member has been counseled to reduce risk of fat-soluble nutrient deficiency by taking daily supplements that contain amounts of vitamin E, alpha-linolenic acid, eicosapentaenoic acid (EHA), and docosahexaenoic acid (DHA) as recommended in the Juxtapid prescribing information
- ❑ The member's dose will **NOT** exceed the age-specific maximum recommended dose

Medication being provided by Specialty Pharmacy – Proprium Rx

Not all drugs may be covered under every Plan

If a drug is non-formulary on a Plan, documentation of medical necessity will be required.

*****Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.*****

****Previous therapies will be verified through pharmacy paid claims or submitted chart notes.****

***Approved by Pharmacy and Therapeutics Committee**: ~~10/17/2013; 5/28/2026;~~ **9/24/2026**
REVISED/UPDATED/REFORMATTED: ~~8/14/2017; 6/18/2019; 6/15/2026;~~ **10/15/2026**