Proprotein Convertase Subtilisin/Kexin type 9 (PCSK9) Inhibitors Prior Authorization with Quantity Limit Program Summary - Through Preferred Agent(s)

This program applies to Medicaid.

Preferred products are:
- **Praluent:**
  - NDC 72733-5901-02
  - NDC 72733-5902-02
- **Repatha:**
  - NDC 72511-0750-01
  - NDC 72511-0760-01
  - NDC 72511-0760-02
  - NDC 72511-0770-01

### FDA INDICATIONS AND DOSING

<table>
<thead>
<tr>
<th>Agents</th>
<th>Indications*</th>
<th>Strength(s)</th>
<th>Dosing and Administration</th>
</tr>
</thead>
</table>
| **Praluent®** (alirocumab) | Adjunctive therapy to diet and maximally tolerated statins for treatment of adults with heterozygous familial hypercholesterolemia (HeFH) or clinical atherosclerotic cardiovascular disease who require additional lowering of LDL cholesterol† | 75 mg/mL pre-filled pen and syringe  
150 mg/mL pre-filled pen and syringe | Atherosclerotic cardiovascular disease or HeFH: 75 mg SC^ once every 2 weeks. May increase dose up to 150 mg SC^ every 2 weeks if the LDL-C response is inadequate±.  
For patients with HeFH undergoing LDL apheresis: 150 mg once every 2 weeks. |
| **Repatha®** (evolocumab) | To reduce the risk of myocardial infarction, stroke, and coronary revascularization in adults with established cardiovascular disease  
Adjunct to diet, alone or in combination with other lipid lowering therapies (e.g., statins, ezetimibe), for treatment of adults with primary hyperlipidemia (including heterozygous familial hypercholesterolemia (HeFH) to reduce low-density lipoprotein cholesterol | 140 mg/mL pre-filled pen and autoinjector  
420 mg/3.5 mL Pushtronex system (infusor with pre-filled cartridge) | Adults with established cardiovascular disease or Primary hyperlipidemia with CVD or HeFH: 140 mg SC^ every 2 weeks or 420 mg SC^ monthly  
HoFH: 420 mg SC^ once monthly |
ezetimibe, LDL apheresis) in patients with homozygous familial hypercholesterolemia (HoFH) who require additional lowering of LDL-C

*Sub-cutaneous

**CLINICAL RATIONALE**

**Heterozygous familial hypercholesterolemia (HeFH)**

Criteria have been developed to aid in diagnosing HeFH. These include the Simon Broome Register criteria and Dutch Lipid clinic Network criteria.\(^6\) Definitive diagnosis of HeFH according to Simon Broome diagnostic criteria requires the patient has one of the following:\(^4\)\(^,\)\(^6\)

- Total cholesterol greater than 6.7 mmol/L or low-density lipoprotein cholesterol (LDL-C) greater than 4.0 mmol/L in a child aged younger than 16 years, or greater than 7.5 mmol/L or LDL-C greater than 4.9 mmol/L in an adult (levels either pre-treatment or highest on treatment) plus tendon xanthomas in the patient, or in first-degree relative (parent, sibling or child), or in second-degree relative (e.g. grandparent, uncle or aunt)

Or

- DNA-based evidence of an LDL receptor mutation, familial defective Apo B-100, or a PCSK9 mutation

The Dutch Lipid Clinic Network criteria assign points based on cholesterol levels, family history of hyperlipidemia or cardiovascular disease, clinical presentation, and/or presence of identified genetic mutation affecting plasma LDL-C.\(^6\)\(^,\)\(^8\) A definitive diagnosis of HeFH can be made in patients with greater than 8 points.

**Dutch Lipid Clinic Network criteria for diagnosis of heterozygous familial hypercholesterolemia**\(^8\)

<table>
<thead>
<tr>
<th>Group 1: Family history</th>
<th>Points</th>
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<tr>
<td>First-degree relative with known premature (≤55 years, men; ≤60 years, women) coronary heart disease (CHD)</td>
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<td>First-degree relative with known LDL cholesterol &gt;95th percentile by age and gender for country</td>
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<td>First-degree relative with tendon xanthoma and/or corneal arcus</td>
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</tr>
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<tr>
<th>Group 2: Clinical history</th>
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</tr>
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<tbody>
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<td>Subject has premature (≤55 years, men; ≤60 years, women) CHD</td>
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<th>Group 5: Molecular genetic testing (DNA analysis)</th>
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<td>Causative mutation shown in the LDLR, APOB, or PCSK9 genes</td>
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**Use and Interpretation**

Assign only one score, the highest applicable, per group then add the points from each group to achieve the total score.

- Definitive FH diagnosis: > 8 points
- Probable FH diagnosis: 6 to 8 points
- Possible FH diagnosis: 3 to 5 points
- Unlikely FH diagnosis: 0 to 2 points
Homozygous familial hypercholesterolemia (HoFH)

Guidelines advise that diagnosis of HoFH can be made on the basis of genetic or clinical criteria. Genetic confirmation of the HoFH includes confirmation of two mutant alleles at the LDL-R, APOB, PCSK9, or LDLRAP1 genes.\textsuperscript{5,6} While genetic testing may provide a definitive diagnosis of HoFH, it is recognized that in some patients, genetic confirmation remains elusive, despite exhaustive investigation; indeed, the existence of additional FH genes cannot be excluded. Historically, HoFH has been most commonly diagnosed on the basis of either an untreated LDL-C plasma concentration $>13$ mmol/L ($>500$ mg/dL), or a treated LDL-C concentration of $\geq 8$ mmol/L ($\geq 300$ mg/dL), accompanied by the presence of cutaneous or tendon xanthomas before the age of 10 years, or the presence of untreated elevated LDL-C levels consistent with HeFH in both parents.\textsuperscript{5,6}

The goal of treatment for FH is to reduce the risk of coronary heart disease (CHD) or risk of a CHD-equivalent condition (e.g. carotid artery disease, diabetes, peripheral arterial disease, or abdominal aortic aneurysm).\textsuperscript{3} According the American Heart Association (AHA), initial treatment for FH should include a high intensity statin.\textsuperscript{9} If the LDL-C is not at goal after 3 months of therapy with the high intensity statin and the patient has been adherent, AHA recommends the addition of ezetimibe. For patients who do not respond to this two drug regimen within 3 months, AHA recommends addition of a PCKS9, a bile acid sequestrant, or niacin. Patients with HoFH who require additional therapy despite treatment with the three drug regimen, AHA recommends addition of Juxtapid or Kynamro and LDL apheresis.\textsuperscript{9}

The AHA/ACC/AACVPR/AAPA/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA Guideline on the Management of Blood Cholesterol states the following regarding PCSK9 therapy\textsuperscript{10}

- Severe hypercholesterolemia (LDL-C $\geq 190$ mg/dL [$\geq 4.9$ mmol/L])
  - In patients 30-75 years of age with heterozygous FH and with an LDL-C level of 100 mg/dL ($\geq 2.6$ mmol/L) or higher while taking maximally tolerated statin and ezetimibe therapy, the addition of a PCSK9 inhibitor may be considered
  - In patients 40-75 years of age with a baseline LDL-C level of 220 mg/dL ($\geq 5.7$ mmol/L) or higher while receiving maximally tolerated statin and ezetimibe therapy, the addition of a PCSK9 inhibitor may be considered

Atherosclerotic Cardiovascular Disease (ASCVD) – Secondary Prevention

The AHA/ACC/AACVPR/AAPA/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA guideline lists the following as clinical ASCVD:

- Acute coronary syndrome (ACS)
- Myocardial infarction (MI)
- Stable or unstable angina or coronary or other arterial revascularization
- Stroke
- Transient ischemic attack (TIA) or peripheral artery disease (PAD) including aortic aneurysm

The AHA/ACC/AACVPR/AAPA/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA Guideline on the Management of Blood Cholesterol states the following regarding PCSK9 therapy:\textsuperscript{10}

- Secondary atherosclerotic cardiovascular disease (ASCVD) prevention
  - In patients with clinical ASCVD who are judged to be very high risk and considered for PCSK9 inhibitor therapy, maximally tolerated LDL-C lowering therapy should include maximally tolerated statin therapy and ezetimibe
  - In patients with clinical ASCVD who are judged to be very high risk and who are on maximally tolerated LDL-C lowering therapy with LDL-C 70 mg/dL ($\geq 1.8$ mmol/L) or higher or a non-HDL-C level of 100 mg/dL ($\geq 2.6$ mmol/L) or higher, it is reasonable to add PCSK9 inhibitor following a clinical-patient discussion about the net benefit, safety, and cost

The AHA/ACC/AACVPR/AAPA/ABC/ACPM/ADA/AGS/APhA/ASPC/NLA/PCNA guideline categorizes the following statin intensities:\textsuperscript{10}

<table>
<thead>
<tr>
<th>Statin Intensity</th>
<th>High Intensity</th>
<th>Moderate Intensity</th>
<th>Low Intensity</th>
</tr>
</thead>
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<table>
<thead>
<tr>
<th>LDL-C Lowering</th>
<th>≥50%</th>
<th>30%-49%</th>
<th>&lt;30%</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Statins</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atorvastatin 40-80 mg</td>
<td>Atorvastatin 10-20 mg</td>
<td>Simvastatin 10 mg</td>
<td></td>
</tr>
<tr>
<td>Rosuvastatin 20-40 mg</td>
<td>Rosuvastatin 5-10 mg</td>
<td>Pravastatin 10-20 mg</td>
<td></td>
</tr>
<tr>
<td>Simvastatin 20-40 mg*</td>
<td>Pravastatin 40-80 mg</td>
<td>Lovastatin 20 mg</td>
<td></td>
</tr>
<tr>
<td>Pravastatin 40-80 mg</td>
<td>Lovastatin 40-80 mg</td>
<td>Fluvastatin 20-40 mg</td>
<td></td>
</tr>
<tr>
<td>Simvastatin 20-40 mg*</td>
<td>Fluvastatin XL 80 mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fluvastatin 40 mg twice daily</td>
<td>Fluvastatin 40 mg twice daily</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pitavastatin 1-4 mg</td>
<td></td>
<td></td>
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</tbody>
</table>

*Although simvastatin 80 mg was evaluated in RCTs, initiation of simvastatin 80 mg or titration to 80 mg is not recommended by the FDA because of the risk of myopathy, including rhabdomyolysis.

**REFERENCES**

Proprotein Convertase Subtilisin/Kexin type 9 (PCSK9) Inhibitors Prior Authorization with Quantity Limit-Through Preferred Agent(s) - Medicaid

TARGET AGENTS
Praluent® (alirocumab)
Repatha® (evolocumab)

PRIOR AUTHORIZATION AND QUANTITY LIMIT TARGET DRUGS- RECOMMENDED LIMITS

<table>
<thead>
<tr>
<th>Brand (generic)</th>
<th>GPI</th>
<th>Multisource Code</th>
<th>Quantity Limit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Praluent (alirocumab)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>75 mg/mL pre-filled pen</td>
<td>3935001000D220</td>
<td>M, N, O, or Y</td>
<td>1 package of 2 pens/28 days</td>
</tr>
<tr>
<td>75 mg/mL pre-filled syringe</td>
<td>3935001000E520</td>
<td>M, N, O, or Y</td>
<td>1 package of 2 syringes/28 days</td>
</tr>
<tr>
<td>150 mg/mL pre-filled pen</td>
<td>3935001000D230</td>
<td>M, N, O, or Y</td>
<td>1 package of 2 pens/28 days</td>
</tr>
<tr>
<td>150 mg/mL pre-filled syringe</td>
<td>3935001000E530</td>
<td>M, N, O, or Y</td>
<td>1 package of 2 syringes/28 days</td>
</tr>
<tr>
<td>Repatha (evolocumab)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>140 mg/mL pre-filled syringe</td>
<td>3935002000E520</td>
<td>M, N, O or Y</td>
<td>2 syringes/28 days</td>
</tr>
<tr>
<td>140 mg/mL pre-filled autoinjector</td>
<td>3935002000D520</td>
<td>M, N, O or Y</td>
<td>2 pens/28 days</td>
</tr>
<tr>
<td>420 mg/3.5 mL single-use Pushtronex system (infusor with pre-filled cartridge)</td>
<td>3935002000E230</td>
<td>M, N, O or Y</td>
<td>1 Pushtronex system/30 days</td>
</tr>
</tbody>
</table>

INITIAL PRIOR AUTHORIZATION CRITERIA FOR APPROVAL
Target Agents will be approved when ALL of the following are met:

1. The patient has ONE of the following:
   A. A diagnosis of heterozygous familial hypercholesterolemia (HeFH) confirmed by ONE of the following:
      i. Genetic confirmation of one mutant allele at the LDLR, Apo-B, PCSK9, or ARH adaptor protein 1/LDLRAP1 gene locus
      OR
      ii. BOTH of the following:
          1. ONE of the following:
              a. History of total cholesterol ≥ 290 mg/dL (>7.5 mmol/L) (pretreatment or highest level while on treatment)
              OR
              b. History of LDL-C ≥ 190 mg/dL (>4.9 mmol/L) (pretreatment or highest level while on treatment)

      AND
   2. History of tendon xanthomas in ONE of the following:
      a. The patient
      OR
      b. The patient’s first degree relative (i.e. parent, sibling, or child)
      OR
      c. The patient’s second degree relative (e.g. grandparent, uncle, or aunt)

      iii. The Patient has a Dutch Lipid Clinic Network Criteria score of greater than 8 (see scoring algorithm in Table 2 below)

      OR
B. A diagnosis of homozygous familial hypercholesterolemia (HoFH) confirmed by ONE of the following:
   i. Genetic confirmation of two mutant alleles at the LDLR, Apo-B, PCSK9, or ARH adaptor protein 1/LDLRAP1 gene locus
      OR
   ii. History of untreated LDL-C >500 mg/dL (>13 mmol/L) or treated LDL-C ≥300 mg/dL (≥7.76 mmol/L) with ONE of the following:
      1. The patient had cutaneous or tendon xanthoma before age 10 years
      OR
      2. Untreated elevated cholesterol levels consistent with heterozygous FH in both parents [untreated LDL-C >190 mg/dL (>4.9 mmol/L) or untreated total cholesterol greater than 290 mg/dL (>7.5 mmol/L)]
      OR
C. BOTH of the following:
   i. A diagnosis of clinical atherosclerotic cardiovascular disease (ASCVD) defined as having ONE of the following:
      1. Acute coronary syndrome
      2. History of myocardial infarction
      3. Stable or unstable angina
      4. Coronary or other arterial revascularization
      5. Stroke
      6. Transient ischemic attack
      7. Peripheral arterial disease, including aortic aneurysm, presumed to be of atherosclerotic origin
      AND
   ii. The patient has very high risk for future ASCVD events
      OR
D. The patient has primary hyperlipidemia AND ALL of the following:
   i. The patient is 40-75 years of age
      AND
   ii. The patient has a LDL-C level ≥ 220 mg/dL (≥5.7 mmol/L) while receiving maximally tolerated statin and ezetimibe therapy
      AND
2. ONE of the following:
   A. The patient is currently adherent* (for the past 90 days) to high-intensity statin therapy (i.e. rosuvastatin 20-40 mg, atorvastatin 40-80 mg, or simvastatin 80 mg)
      OR
   B. BOTH of the following:
      i. The patient has tried and is intolerant* to high-intensity statin therapy (i.e. rosuvastatin 20-40 mg and atorvastatin 40-80mg)
      AND
      ii. The patient is currently adherent* (for the past 90 days) to low or moderate intensity statin therapy
      OR
   C. The patient has documented intolerance* to TWO different statins
      OR
   D. The patient has an FDA labeled contraindication to a statin
      AND
3. ONE of the following:
   A. The patient has not achieved a 50% reduction in LDL-C from baseline while on a maximally tolerated statin
      OR
   B. The patient has an LDL-C ≥70 mg/dL (≥1.81 mmol/L) evaluated within the past 90 days
      OR
   C. The patient has ASCVD AND a non-HDL-C level of ≥100 mg/dL (≥2.6 mmol/L) evaluated within the past 90 days
      AND
4. If the client has a preferred agent, ONE of the following:
   A. The requested agent is the preferred agent
      OR
   B. ONE of the following:
      i. The patient has tried and had an inadequate response to the preferred agent
         OR
      ii. The patient has a documented intolerance, FDA labeled contraindication, or
           hypersensitivity to the preferred agent that is not expected to occur with the
           requested agent
   AND
5. ONE of the following:
   A. The patient is not currently taking another PSCK9 agent
      OR
   B. The other PCSK9 agent will be discontinued before starting therapy with the requested
      agent
   AND
6. The agent is being prescribed by a specialist in the area of practice related to the patient’s
   diagnosis (e.g. cardiologist, endocrinologist, or lipid specialist) or in consultation with a specialist in
   the area of practice related to the patient’s diagnosis
   AND
7. The patient does not have any FDA labeled contraindication(s) to therapy with the requested agent
   AND
8. ONE of the following:
   A. The requested quantity (dose) does not exceed the program quantity limit
      OR
   B. ALL of the following:
      i. The requested quantity (dose) is greater than the program quantity limit
         AND
      ii. The requested quantity (dose) does not exceed the maximum FDA labeled dose (for
          the requested indication)
         AND
      iii. The requested quantity (dose) cannot be achieved with a lower quantity of a higher
           strength that does not exceed the program quantity limit

^Adherence is defined as filling ≥80% of therapy as prescribed in the past 90 days
*intolerance is defined as inability to tolerate the lowest FDA approved starting dose of a statin

Length of Approval: 12 months

Renewal Evaluation
Target Agents will be approved for renewal when ALL of the following criteria are met:
1. The patient has been previously approved for therapy for PCSK9 inhibitors through Prime
   Therapeutics PA process
   AND
2. If the client has a preferred agent, ONE of the following:
   a. The requested agent is the preferred agent
      OR
   b. ONE of the following:
      i. The patient has tried and had an inadequate response to the preferred agent
         OR
      ii. The patient has a documented intolerance, FDA labeled contraindication, or
          hypersensitivity to the preferred agent that is not expected to occur with the
          requested agent
   AND
3. The patient has shown clinical benefit with the requested agent
   AND
4. The patient is currently adherent^ (for the past 90 days) to therapy with the requested agent
AND
5. ONE of the following:
   a. The patient is currently adherent\(^\text{^}\) (for the past 90 days) to high-intensity statin therapy (i.e. rosuvastatin 20-40 mg, atorvastatin 40-80 mg, or simvastatin 80 mg)
   \textbf{OR}
   b. BOTH of the following:
      i. The patient has tried and is intolerant\(^*\) to high-intensity statin therapy (i.e. rosuvastatin 20-40 mg and atorvastatin 40-80 mg) \textbf{AND}
      ii. The patient is currently adherent\(^\text{^}\) (for the past 90 days) to low or moderate intensity statin therapy
   \textbf{OR}
   c. The patient has documented intolerance\(^*\) to TWO different statins
   \textbf{OR}
   d. The patient has an FDA labeled contraindication to a statin
   \textbf{AND}
6. ONE of the following:
   a. The patient is not currently taking another PCSK9 agent
   \textbf{OR}
   b. The patient will discontinue the current PCSK9 agent before starting therapy with the requested agent
   \textbf{AND}
7. The agent is being prescribed by a specialist in the area of practice related to the patient’s diagnosis (e.g. cardiologist, endocrinologist, lipid specialist) or in consultation with a specialist in the area of practice related to the patient’s diagnosis
   \textbf{AND}
8. The patient does not have any FDA labeled contraindication(s) to therapy with the requested agent
   \textbf{AND}
9. ONE of the following:
   a. The requested quantity (dose) does not exceed the program quantity limit
   \textbf{OR}
   b. ALL of the following:
      i. The requested quantity (dose) is greater than the program quantity limit \textbf{AND}
      ii. The requested quantity (dose) does not exceed the maximum FDA labeled dose (for the requested indication) \textbf{AND}
      iii. The requested quantity (dose) cannot be achieved with a lower quantity of a higher strength that does not exceed the program quantity limit

\(^\text{^}\)Adherence is defined as filling ≥80% of therapy as prescribed in the past 90 days
\(^*\)intolerance is defined as inability to tolerate the lowest FDA approved starting dose of a statin

\textbf{Length of approval: 12 months}

\textbf{Table 2: Dutch Lipid Clinic Network criteria for diagnosis of heterozygous familial hypercholesterolemia}\(^\text{23}\)

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