Evinacumab (Evkeeza) – PA Criteria

HCPC: J1305

Evkeeza (evinacumab), an angiopoietin-like 3 (ANGPTL3) inhibitor monoclonal antibody approved as an adjunct to other low-density lipoprotein-cholesterol (LDL-C) lowering therapies for the treatment of individuals with homozygous familial hypercholesterolemia (HoFH). It is covered by South Dakota Medicaid following prior authorization when the patient meets the following criteria:

- **Initial Therapy (must meet all):**
  - Therapy is prescribed by or in consultation with a cardiologist, endocrinologist, or lipid specialist
  - Individual has a diagnosis of HoFH defined as **one** of the following:
    - Genetic mutation indicating HoFH (e.g., mutations in low density lipoprotein receptor [LDLR] gene, proprotein convertase subtilisin kexin 9 [PCSK9] gene, apolipoprotein B [apo B] gene, low density lipoprotein receptor adaptor protein 1 [LDLRAP1] gene)
    - Treated LDL-C ≥ 300 mg/dL and one of the following
      - Tendinous or cutaneous xanthoma prior to age 10 years
      - Evidence of HeFH in both parents (e.g., documented history of elevated LDLC ≥ 190 mg/dL prior to lipid-lowering therapy)
    - Untreated LDL-C ≥ 500 mg/dL
  - Individual meets **one** of the following regarding statin therapy
    - Individual is currently taking high intensity statin therapy (high intensity statin is defined at atorvastatin ≥40mg or rosuvastatin ≥20mg) and has been compliant with therapy for ≥3 months
    - Individual is statin intolerant based on **one** of the following
      - Inability to tolerate at least two statins, with at least one started at the lowest daily starting dose
      - Development of statin associated rhabdomyolysis or immune-mediated necrotizing myopathy (IMNM) after a trial of one statin
    - Individual has a contraindication to statin therapy including but not limited to active liver disease, unexplained persistent elevation of hepatic transaminases or pregnancy
  - Individual meets **one** of the following regarding ezetimibe therapy
    - Individual is currently taking at a dose of 10mg daily and has been compliant with therapy for ≥3 months
    - Individual has had a trial and inadequate response to ezetimibe therapy
    - Individual is intolerant of ezetimibe as documented by provider
  - Individual meets **one** of the following regarding proprotein convertase subtilisin kexin type 9 (PCSK9) inhibitor therapy
    - Individual is currently taking PCSK9 therapy with evolocumab (Repatha) or alirocumab (Praluent) at FDA approved doses and has been compliant with therapy for ≥3 months.
    - Individual has had a trial and inadequate response to PCSK9 therapy as indicated by LDL reduction of ≤50% from baseline
    - Individual is intolerant of PCSK9 therapy
    - Documentation is provided that individual is LDLR negative
  - Therapy will not be used concomitantly with lomitapide (Juxtapid)
  - Individual is ≥5 years of age
  - Approval duration: 6 months
- **Continuation of Therapy:**
  - Individual continues to meet initial criteria
  - Individual continues to use in combination with other lipid lowering therapies including maximum tolerated statin, ezetimibe and PCSK9 inhibitor therapy (unless previously waived due to contraindication or intolerance)

- Documentation is provided individual has had a reduction in total LDL-C since starting Evkeeza therapy
- Approval duration: 1 year