Vyndamax (tafamidis)

Meets primary Coverage Criteria Or Is Covered For Contracts Without Primary Coverage Criteria

Tafamidis/Tafamidis Meglumine meets member benefit certificate primary coverage criteria that there be scientific evidence of effectiveness in improving health outcomes for the treatment of cardiomyopathy of wild type or hereditary transthyretin-mediated amyloidosis in adults to reduce cardiovascular mortality and cardiovascular-related hospitalization when **ALL** the following are met:

INITIAL APPROVAL for 6 months:

1. Diagnosis of cardiomyopathy due to hereditary transthyretin amyloidosis (hATTR) OR wild type transthyretin amyloidosis (ATTRwt) – documentation from the medical record must be provided in the form of one of the following (Maurer, 2018):

    a. Presence of amyloid deposits in biopsy tissue – laboratory documentation must be provided; **OR**

    b. Presence of a variant TTR genotype and/or TTR precursor protein identification by:

        i. Immunohistochemistry or mass spectrometry of a biopsy specimen; **OR**

        ii. Positive scintigraphy (99m Tc-pyrophosphate), with a *semi-quantitative visual score (Myocardial Uptake by Comparison to Bone of 2 or 3, or Heart-to-Contralateral Lung Ratio (H/CL) of >1.5.

*Semi –quantitative visual grading of myocardia 99m Tc-PYP Uptake by Comparison to Bone (rib) Uptake:

Grade 0—no myocardial uptake and normal rib uptake

Grade 1—myocardial uptake less than rib uptake

Grade 2—myocardial uptake equal to rib uptake

Grade 3—myocardial uptake greater than rib uptake with mild/absent rib uptake; **AND**

2. Current or historical evidence for NYHA Class I, II, or III heart failure (HF), including either (Maurer, 2018):

    a. At least 1 prior hospitalization for HF with clinical evidence of HF manifested by signs or symptoms of:

        i. Volume overload (Maurer, 2018); **OR**

        ii. Elevated intracardiac pressures (e.g., elevated jugular venous pressure, shortness of breath or signs of pulmonary congestion on x-ray or auscultation, peripheral edema) that required/requires treatment with a diuretic for

improvement)(Maurer, 2018) – documentation from the medical record must be provided; **OR**

b. Submission of lab report showing NT-pro BNP > 600 (Maurer, 2018); **AND**

3. Submission of Echocardiogram results demonstrating an End-diastolic interventricular septal wall thickness is greater than 12 mm – documentation from the medical record must be provided (Maurer, 2018); **AND**

4. Primary (light chain) amyloidosis has been excluded by serum and urine immunofixation and serum free light chain assay (Maurer, 2018); **AND**

5. Must not be taking concurrently with inotersen or patisiran; **AND**

6. Individual does not have any of the following conditions:

a. Class 4 heart failure; **OR**

b. Prior liver transplantation; **OR**

c. Prior heart transplantation; **OR**

d. In combination with inotersen (e.g., Tegsedi) or patisiran (e.g., Onpattro); **AND**

7. Must be dosed in accordance with the FDA label.

CONTINUED APPROVAL:

1. Prior approval by Arkansas Blue Cross in the past two years, or the member currently meets all the coverage criteria.

2. Description of a clinically meaningful beneficial response to treatment with tafamidis such as improvement or slowing of progression of exercise intolerance, BNP, or fluid overload compared to baseline – documentation from the medical record must be provided.

3. Submission of current results of NT-pro BNP.

Dosage and Administration

Dosing per FDA Guidelines

The recommended dosage of Tafamidis (e.g., Vyndamax) is 61 mg (one capsule) once daily.

The recommended dosage of Tafamidis Meglumine (e.g., Vyndaqel) is 80 mg (four 20mg capsules) daily.

Tafamidis meglumine is available as 20 mg tablets and tafamidis is available as 61 mg.

Please refer to a separate policy on Site of Care or Site of Service Review (policy #2018030) for pharmacologic/biologic medications.

Does Not Meet Primary Coverage Criteria Or Is Investigational For Contracts Without Primary Coverage Criteria

The use of Tafamidis or Tafamidis Meglumine, for any indication or circumstance not described above, does not meet primary certificate coverage criteria that there be scientific evidence of effectiveness in improving health outcomes.

For members with contracts without primary coverage criteria, Tafamidis or Tafamidis Meglumine, for any indication or circumstance not described above, is considered **investigational**.

Investigational services are specific to contract exclusions in most member benefit certificates of coverage.