VENTURA COUNTY
HEALTH CARE PLAN

PRIOR AUTHORIZATION POLICY

POLICY: Amyloidosis – Acoramidis Prior Authorization Policy
- Attruby (acoramidis tablets – Bridgebio)

REVIEW DATE: 12/04/2024

OVERVIEW

Attruby, a transthyretin (TTR) stabilizer, is indicated for the treatment of the **cardiomyopathy of wild-type or variant TRR-mediated amyloidosis (ATTR-CM)** to reduce cardiovascular death and cardiovascular-related hospitalization in adults.[1] Studies excluded patients with New York Heart Association class IV disease.[2]

Disease Overview

ATTR-CM is a progressive, debilitating, and life-threatening disease that may go unrecognized and result in late-stage diagnosis.[3,4] ATTR can be caused by an acquired wild-type variant or be an inherited variant that alters the structure and function of TTR. Genetic testing of the TTR gene is done to differentiate patients with the wild-type vs. a genetic variant. Although patients with ATTR may present with a variety of symptoms, neuropathy or cardiomyopathy are often the most prominent symptoms. Patients may also present with a mixed phenotype and exhibit signs of both neuropathy and cardiomyopathy. In ATTR-CM, the stability of the TTR tetramer is altered, resulting in misfolding of the TTR protein. This leads to accumulation of amyloid in the body, most commonly the myocardium and peripheral nerves. As a result of amyloid deposits in cardiac tissue and causes the heart muscle becomes stiff and rigid. Although the clinical course varies, patients typically exhibit heart failure with preserved ejection fraction. However, some patients may present with right-sided heart failure, atrial fibrillation, aortic stenosis and complete heart block, angina, or cardiogenic shock.

Guidelines

Attruby is currently not addressed in guidelines. The American Heart Association (AHA) scientific statement for the evolving diagnosis and management of cardiac amyloidosis (2020) recognizes tafamidis as a treatment for ATTR-CM.[5] They note that the benefit of tafamidis has not been observed in patients with NYHA class IV symptoms. Additionally, although combination use of tafamidis with Onpattro® (patisiran lipid complex intravenous infusion) or Tegsedi® (inotersen subcutaneous injection) is appealing to target both TTR silencing and stabilization for the remaining synthesized protein, this approach lacks data and may be cost-prohibitive. Tafamidis should generally be considered the agent of choice in ATTR-CM in patients with reasonable expected survival according to a position statement of the European Society of Cardiology (ESC) working group on myocardial and pericardial disease (2021).[6] The working group notes that tafamidis is the only drug that has shown efficacy in a randomized trial in patients with ATTR-CM and should be considered in patients with reasonable expected survival. The American College of Cardiology (ACC) expert consensus decision pathway on comprehensive multidisciplinary care for patients with cardiac amyloidosis (2023) make similar comments and recommendations to the AHA and ESC regarding tafamidis.[7]

POLICY STATEMENT

Prior Authorization is recommended for prescription benefit coverage of Attruby. All approvals are provided for the duration noted below. Because of the specialized skills required for evaluation and diagnosis of patients treated with Attruby as well as the monitoring required for adverse events and long-

term efficacy, initial approval requires the agent to be prescribed by or in consultation with a physician who specializes in the condition being treated.

Automation: None.

RECOMMENDED AUTHORIZATION CRITERIA

Coverage of Attruby is recommended in those who meet the following criteria:

FDA-Approved Indication

1. **Cardiomyopathy of Wild-Type or Hereditary Transthyretin-Mediated Amyloidosis (ATTR-CM).** Approve for 1 year if the patient meets ALL of the following (A, B, C, D, and E):
   Note: Variant Transthyretin Amyloidosis is also known as Hereditary Transthyretin Amyloidosis.
   - **A)** Patient is ≥ 18 years of age; AND
   - **B)** The diagnosis was confirmed by ONE of the following (i, ii, or iii):
     - **i.** A technetium pyrophosphate scan (i.e., nuclear scintigraphy); OR
     - **ii.** A tissue biopsy with confirmatory transthyretin (TTR) amyloid typing by mass spectrometry, immunoelectron microscopy or immunohistochemistry; OR
     - **iii.** Patient had genetic testing which, according to the prescriber, identified a transthyretin (TTR) pathogenic variant; AND
       Note: Examples of TTR variants include Val122Ile variant and Thr60Ala variant. If the patient has wild-type amyloidosis, this is **not** a TTR pathogenic variant.
   - **C)** Diagnostic cardiac imaging has demonstrated cardiac involvement; AND
     Note: Examples of cardiac imaging include echocardiogram and cardiac magnetic imaging. Examples of cardiac involvement on imaging include increased thickness of the ventricular wall or interventricular septum.
   - **D)** Patient has heart failure, but does **not** have New York Heart Association class IV disease; AND
   - **E)** The medication is prescribed by or in consultation with a cardiologist or a physician who specializes in the treatment of amyloidosis.

CONDITIONS NOT RECOMMENDED FOR APPROVAL

Coverage of Attruby is not recommended in the following situations:

1. **Concurrent use with other medications indicated for the treatment of polyneuropathy of hereditary transthyretin-mediated amyloidosis or transthyretin-mediated amyloidosis-cardiomyopathy (e.g., Amvuttra [vutrisiran subcutaneous injection], Onpattro [patisiran intravenous infusion], Tegsedi [inotersen subcutaneous injection], Wainua [eplontersen subcutaneous injection], or a tafamidis product).**
   The requested medication should not be administered in combination with other medications indicated for polyneuropathy of hereditary transthyretin-mediated amyloidosis or transthyretin-mediated amyloidosis-cardiomyopathy. Combination therapy is generally not recommended due to a lack of controlled clinical trial data supporting additive efficacy.

2. **Polyneuropathy of Hereditary Transthyretin–Mediated Amyloidosis (hATTR).** Attruby is not indicated for treatment of symptoms of polyneuropathy associated with hATTR.[1]
   Note: For patients with hATTR and cardiomyopathy or mixed phenotype (concurrent cardiomyopathy and polyneuropathy), refer to FDA-Approved Indication, above.

3. Coverage is not recommended for circumstances not listed in the Recommended Authorization Criteria. Criteria will be updated as new published data are available.

REFERENCES

1. Attruby tablets [prescribing information]. Palo Alto, CA: BridgeBio; November 2024.
2. Gillmore JD, Judge DP, Cappelli F, et al. Efficacy and safety of acoramidis in transthyretin amyloid cardiomyopathy. NEJM. 2024;390(2):132-142.
3. National Library of Medicine, StatPearls. Transthyretin Amyloid Cardiomyopathy (ATTR-CM). Available at. https://www.ncbi.nlm.nih.gov/books/NBK574531/. Accessed November 11, 2024.
4. Vaishnav J, Brown E, Sharma K. Advances in the diagnosis and treatment of transthyretin amyloid cardiomyopathy. *Prog Cardiovase Dis.* 2024;82:113-124.
5. Kittleson MM, Maurer MS, Ambardekar AV, et al; on behalf of the American Heart Association Heart Failure and Transplantation Committee of the Council on Clinical Cardiology. AHA scientific statement: cardiac amyloidosis: evolving diagnosis and management. *Circulation.* 2020;142:e7-e22.
6. Garcia-Pavia P, Rapezzi C, Adler Y, et al. Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC working group on myocardial and pericardial disease. *Eur Heart J.* 2021;42:1554-1568.
7. Kittleson M, Ruberg FL, Ambardekar AV, et al. A report of the American College of Cardiology Solution Set Oversight Committee. 2023 ACC expert consensus decision pathway on comprehensive multidisciplinary care for the patient with cardiac amyloidosis. *JACC.* 2023;81(11):1076-1126.

HISTORY

Type of Revision	Summary of Changes	Review Date
New Policy	--	12/04/2024