CVS Caremark®

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| Reference number(s) |
| 3076-A |

# Specialty Guideline Management Vyndaqel-Vyndamax

## Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

| Brand Name | Generic Name |
| --- | --- |
| Vyndaqel | tafamidis meglumine |
| Vyndamax | tafamidis |

## Indications

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

### FDA-approved Indication1

Vyndaqel and Vyndamax are transthyretin stabilizers indicated for the treatment of the cardiomyopathy of wild-type or hereditary transthyretin-mediated amyloidosis (ATTR-CM) in adults to reduce cardiovascular mortality and cardiovascular-related hospitalization.

All other indications are considered experimental/investigational and not medically necessary.

## Documentation

Submission of the following information is necessary to initiate the prior authorization review:

### Initial requests

* Chart notes or medical record documentation confirming the member demonstrates clinical symptoms of cardiomyopathy and heart failure
* For biopsy proven disease:
  + Tissue biopsy confirming the presence of the transthyretin amyloid deposition
  + Immunohistochemical analysis, mass spectrometry, tissue staining, or polarized light microscopy results confirming transthyretin precursor proteins
* For technetium-labeled bone scintigraphy proven disease:
  + Scintigraphy tracing results confirming presence of amyloid deposits
  + A serum kappa/lambda free light chain ratio, serum protein immunofixation or urine protein immunofixation test result showing the absence of monoclonal proteins
* For hereditary ATTR-CM: results confirming a mutation of the transthyretin (TTR) gene

### Continuation requests

Chart notes or medical record documentation confirming the member demonstrates a beneficial response to treatment (e.g., improvement in rate of disease progression as demonstrated by distance walked on the 6-minute walk test, the Kansas City Cardiomyopathy Questionnaire–Overall Summary [KCCQ-OS] score, cardiovascular-related hospitalizations, New York Heart Association [NYHA] classification of heart failure, left ventricular stroke volume, N-terminal B-type natriuretic peptide [NT-proBNP] level)

## Coverage Criteria

### Cardiomyopathy of wild-type or hereditary transthyretin-mediated amyloidosis1-5

Authorization of 12 months may be granted for treatment of cardiomyopathy of wild-type or hereditary transthyretin-mediated amyloidosis (ATTR-CM) when all of the following criteria are met:

* The member exhibits clinical symptoms of cardiomyopathy and heart failure (e.g., dyspnea, fatigue, orthostatic hypotension, syncope, peripheral edema).
* The diagnosis is confirmed by either of the following criteria:
* The member meets both of the following criteria:
  + Presence of transthyretin amyloid deposits on analysis of biopsy from cardiac or noncardiac sites.
  + Presence of transthyretin precursor proteins was confirmed by immunohistochemical analysis, mass spectrometry, tissue staining, or polarized light microscopy.
* The member meets both of the following criteria:
  + Positive technetium-labeled bone scintigraphy tracing.
  + Systemic light chain amyloidosis is ruled out by a test showing absence of monoclonal proteins (serum kappa/lambda free light chain ratio, serum protein immunofixation, or urine protein immunofixation).
* For members with hereditary ATTR-CM, presence of a mutation of the TTR gene was confirmed.
* The member is not a liver transplant recipient.
* The requested medication will not be used in combination with inotersen (Tegsedi), patisiran (Onpattro), vutrisiran (Amvuttra), eplontersen (Wainua), or acoramidis (Attruby).

## Continuation of Therapy

Authorization of 12 months may be granted for the continued treatment of ATTR-CM when both of the following criteria are met:

* The member must meet all requirements in the coverage criteria section.
* The member must have demonstrated a beneficial response to treatment with tafamidis therapy (e.g., improvement in rate of disease progression as demonstrated by distance walked on the 6-minute walk test, the Kansas City Cardiomyopathy Questionnaire–Overall Summary [KCCQ-OS] score, cardiovascular-related hospitalizations, New York Heart Association [NYHA] classification of heart failure, left ventricular stroke volume, N-terminal B-type natriuretic peptide [NT-proBNP] level).

## References

1. Vyndaqel and Vyndamax [package insert]. New York, NY: Pfizer Labs; October 2023.
2. Maurer MS, Schwartz JH, Gundapaneni B, et al. Tafamidis treatment for patients with transthyretin amyloid cardiomyopathy. N Engl J Med. 2018;379(11):1007-1016.
3. Maurer MS, Sabahat B, Thibaud D, et al. Expert consensus recommendations for the suspicion and diagnosis of transthyretin cardiac amyloidosis. Circ Heart Fail. 2019;12(9):e006075.
4. Ruberg FL, Grogan M, Hanna M, et al. Transthyretin amyloid cardiomyopathy: JACC State-of-the-Art Review. J Am Coll Cardiol. 2019;73(22):2872-2891.
5. Yadav JD, Othee H, Chan KA, et al. Transthyretin Amyloid Cardiomyopathy-Current and Future Therapies. Ann Pharmacother. 2021;55(12):1502-1514.