



An Independent Licensee of the Blue Cross Blue Shield Association

PHARMACY COVERAGE GUIDELINES  
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 2/21/2019  
LAST REVIEW DATE: 8/19/2021  
LAST CRITERIA REVISION DATE: 8/19/2021  
ARCHIVE DATE:

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

Coverage for services, procedures, medical devices and drugs are dependent upon benefit eligibility as outlined in the member's specific benefit plan. This Pharmacy Coverage Guideline must be read in its entirety to determine coverage eligibility, if any.

This Pharmacy Coverage Guideline provides information related to coverage determinations only and does not imply that a service or treatment is clinically appropriate or inappropriate. The provider and the member are responsible for all decisions regarding the appropriateness of care. Providers should provide BCBSAZ complete medical rationale when requesting any exceptions to these guidelines.

The section identified as "Description" defines or describes a service, procedure, medical device or drug and is in no way intended as a statement of medical necessity and/or coverage.

The section identified as "Criteria" defines criteria to determine whether a service, procedure, medical device or drug is considered medically necessary or experimental or investigational.

State or federal mandates, e.g., FEP program, may dictate that any drug, device or biological product approved by the U.S. Food and Drug Administration (FDA) may not be considered experimental or investigational and thus the drug, device or biological product may be assessed only on the basis of medical necessity.

Pharmacy Coverage Guidelines are subject to change as new information becomes available.

For purposes of this Pharmacy Coverage Guideline, the terms "experimental" and "investigational" are considered to be interchangeable.

BLUE CROSS®, BLUE SHIELD® and the Cross and Shield Symbols are registered service marks of the Blue Cross and Blue Shield Association, an association of independent Blue Cross and Blue Shield Plans. All other trademarks and service marks contained in this guideline are the property of their respective owners, which are not affiliated with BCBSAZ.

---

This Pharmacy Coverage Guideline does not apply to FEP or other states' Blues Plans.

Information about medications that require precertification is available at [www.azblue.com/pharmacy](http://www.azblue.com/pharmacy).

Some large (100+) benefit plan groups may customize certain benefits, including adding or deleting precertification requirements.

All applicable benefit plan provisions apply, e.g., waiting periods, limitations, exclusions, waivers and benefit maximums.

Precertification for medication(s) or product(s) indicated in this guideline requires completion of the [request form](#) in its entirety with the chart notes as documentation. **All requested data must be provided.** Once completed the form must be signed by the prescribing provider and faxed back to BCBSAZ Pharmacy Management at (602) 864-3126 or emailed to [Pharmacyprecert@azblue.com](mailto:Pharmacyprecert@azblue.com). **Incomplete forms or forms without the chart notes will be returned.**

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

**Tegsedi (inotersen)**

**Criteria:**

- **Criteria for initial therapy:** Tegsedi (inotersen) is considered *medically necessary* and will be approved when **ALL** of the following criteria are met:
1. Prescriber is a physician specializing in the patient's diagnosis or is in consultation with a Neurologist
  2. Individual is 18 years of age or older
  3. A confirmed diagnosis of **polyneuropathy of hereditary transthyretin (hTTR)-mediated amyloidosis** and **ALL** of the following:
    - a. Diagnosis confirmed by biopsy **or** genetic testing documenting pathogenic TTR mutation
    - b. Signs and symptoms of polyneuropathy
    - c. Polyneuropathy disability stage III B or lower **OR** Familial amyloid polyneuropathy stage I or II
  4. Individual has failure, contraindication or intolerance to **ALL** the following:
    - a. Diflunisal 250 mg twice daily used continuously and concurrently with a proton pump inhibitor (lansoprazole, omeprazole, pantoprazole, or rabeprazole) for at least 6 months
    - b. The patient has tried or is currently receiving at least **one** systemic agent for symptoms of polyneuropathy from **one** of the following pharmacologic classes: a gabapentin-type product (e.g., gabapentin [Neurontin], Lyrica [pregabalin]) duloxetine, or a tricyclic antidepressant (e.g., amitriptyline, nortriptyline)
  5. **ALL** of the following baseline tests have been completed before initiation of treatment with continued monitoring as clinically appropriate:
    - a. Platelet count is at least  $100 \times 10^9/L$  [Note: This is waved if it is verified that Provider, Patient, and Pharmacy are enrolled in the REMS]
    - b. Serum creatinine [Note: This is waved if it is verified that Provider, Patient, and Pharmacy are enrolled in the REMS]
    - c. eGFR is at least  $45 \text{ mL/min}/1.73 \text{ m}^2$  [Note: This is waved if it is verified that Provider, Patient, and Pharmacy are enrolled in the REMS]
    - d. Urinary protein to creatinine ratio (UPCR)  $< 1,000 \text{ mg/g}$  [Note: This is waved if it is verified that Provider, Patient, and Pharmacy are enrolled in the REMS]
    - e. Urinalysis [Note: This is waved if it is verified that Provider, Patient, and Pharmacy are enrolled in the REMS]
    - f. Alanine aminotransferase (ALT), aspartate aminotransferase (AST), and total bilirubin

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

6. There are **NO** FDA-label contraindications, such as:
  - a. Platelet count < 100 x 10<sup>9</sup>/L
  - b. History of glomerulonephritis caused by Tegsedi
7. Neuropathy is not due to other causes such as from diabetes mellitus, chronic alcohol, vitamin B12 deficiency, chronic inflammatory demyelinating polyneuropathy
8. Will not be used with Onpattro (patisiran), Vyndaqel (tafamidis meglumine), or Vyndamax (tafamidis)
9. Individual does not have moderate or severe hepatic impairment
10. Individual does not have a liver transplant

**Initial approval duration:** One infusion every week for 6 months

➤ **Criteria for continuation of coverage (renewal request):** Tegsedi (inotersen) is considered **medically necessary** and will be approved when **ALL** of the following criteria are met:

1. Individual continues to be seen by a physician specializing in the patient's diagnosis or is in consultation with a Neurologist
2. Individual's condition responded while on therapy
  - a. Response is defined as:
    - i. Achieved and maintains at least a 25% improvement in neurologic function (cranial nerves, reflexes, sensations), motor function (muscle strength), cardiac function (heart rate response to deep breathing, postural blood pressure), quantitative sensory testing (touch-pressure and heat-pain) and peripheral nerve electrophysiology
3. Individual has been adherent with the medication
4. Individual has not developed any contraindications or other significant adverse drug effects that may exclude continued use
  - a. Contraindications as listed in the criteria for initial therapy section
  - b. Significant adverse effect such as:
    - i. Thrombocytopenia
    - ii. Glomerulonephritis
    - iii. Nephrotic syndrome
    - iv. Stroke
    - v. Carotid artery dissection
    - vi. Cytokine release symptoms
    - vii. Hepatic dysfunction
    - viii. Detection of treatment emergent anti-platelet IgG antibodies

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYنداQEL® (tafamidis meglumine) oral**

---

5. Will not be used with Onpattro (patisiran), Vyndaqel (tafamidis meglumine), or Vyndamax (tafamidis)
6. Individual does not have moderate or severe hepatic impairment
7. Individual does not have a liver transplant

**Renewal duration:** One infusion every week for 12 months

➤ Criteria for a request for non-FDA use or indication, treatment with dosing, frequency, or duration outside the FDA-approved dosing, frequency, and duration, refer to one of the following Pharmacy Coverage Guideline:

1. **Off-Label Use of a Non-cancer Medications**
2. **Off-Label Use of a Cancer Medication for the Treatment of Cancer without a Specific Coverage Guideline**

---

**Vyndamax (tafamidis)**  
**Vyndaqel (tafamidis meglumine)**

**Criteria:**

- **Criteria for initial therapy:** Vyndamax (tafamidis) or Vyndaqel (tafamidis meglumine) is considered **medically necessary** and will be approved when **ALL** of the following criteria are met:
1. Prescriber is a physician specializing in the patient's diagnosis or is in consultation with a Cardiologist
  2. Individual is 18 years of age or older
  3. A confirmed diagnosis of symptomatic cardiomyopathy of wild-type or hereditary **transthyretin (TTR)-mediated amyloid** (ATTR-CM) and **ALL** of the following:
    - a. Has a history of at least **one** prior hospitalization for heart failure **or** clinical evidence of heart failure (without hospitalization) requiring diuretics
    - b. Presence of amyloid deposits in tissue biopsy or technetium-based pyrophosphate bone imaging
    - c. There is evidence of amyloid cardiac involvement by echocardiography or cardiac magnetic resonance imaging (e.g., end-diastolic interventricular septal wall thickness of > 12 mm or increased thickness of ventricular wall)
  4. **ONE** of the following:
    - a. **For wild-type ATTR-CM:** immunohistochemical analysis, scintigraphy, or mass spectrometry confirming presence of transthyretin precursor proteins

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

- b. **For hereditary ATTR-CM:** genetic testing confirming TTR gene mutation is pathogenic or likely pathogenic variant
- 5. **ALL** of the following baseline tests have been completed before initiation of treatment with continued monitoring as clinically appropriate:
  - a. 6-minute walk test of > 100 meters
  - b. Plasma NT-proBNP is  $\geq$  600 pg/mL
- 6. Individual does not have heart failure due to other causes other than ATTR
- 7. Individual does not have light chain amyloidosis
- 8. Individual does not have an implanted cardiac device
- 9. Individual does not have symptoms suggestive NYHA functional class III or IV
- 10. Individual does not have liver or heart transplantation
- 11. Individual does not use any non-steroidal anti-inflammatory agent, verapamil, diltiazem, digitalis, doxycycline, or tauroudeoxycholic acid product
- 12. Individual does not have severe hepatic impairment (Child-Pugh Class C)
- 13. Will not be used with Onpattro (patisiran) or Tegsedi (inotersen) or diflunisal
- 14. Vyndamax (tafamidis) and Vyndaqel (tafamidis meglumine) will not be used concurrently or interchangeably

**Initial approval duration:** 6 months

- **Criteria for continuation of coverage (renewal request):** Vyndamax (tafamidis) or Vyndaqel (tafamidis meglumine) is considered **medically necessary** and will be approved when **ALL** of the following criteria are met:
- 1. Individual continues to be seen by a physician specializing in the patient's diagnosis or is in consultation with a Cardiologist
  - 2. Individual's condition responded while on therapy
    - a. Response is defined as:
      - i. Achieved and maintains
        - 1. No worsening of 6-minute walk test of > 100 meters from baseline
        - 2. No worsening of NYHA functional class from baseline
        - 3. Reduction in plasma NT-proBNP from baseline
        - 4. At least a 30% reduction in non-elective cardiovascular related hospitalization

PHARMACY COVERAGE GUIDELINES  
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 2/21/2019  
LAST REVIEW DATE: 8/19/2021  
LAST CRITERIA REVISION DATE: 8/19/2021  
ARCHIVE DATE:

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

3. Individual has been adherent with the medication
4. Individual has not developed any significant level 4 adverse drug effects that may exclude continued use
5. Individual does not have symptoms suggestive NYHA functional class III or IV
6. Individual does not use any non-steroidal anti-inflammatory agent, verapamil, diltiazem, digitalis, doxycycline, or taurourodeoxycholic acid product
7. Individual does not have severe hepatic impairment (Child-Pugh Class C)
8. Will not be used with Onpattro (patisiran) or Tegsedi (inotersen) or diflunisal
9. Vyndamax (tafamidis) and Vyndaqel (tafamidis meglumine) will not be used concurrently or interchangeably
10. There are no significant interacting drugs

**Renewal duration:** 12 months

- Criteria for a request for non-FDA use or indication, treatment with dosing, frequency, or duration outside the FDA-approved dosing, frequency, and duration, refer to one of the following Pharmacy Coverage Guideline:
3. **Off-Label Use of a Non-cancer Medications**
  4. **Off-Label Use of a Cancer Medication for the Treatment of Cancer without a Specific Coverage Guideline**

---

**Description:**

Tegsedi (inotersen) is indicated for the treatment of the polyneuropathy of hereditary transthyretin-mediated amyloidosis (hATTR amyloidosis) in adults.

Tegsedi (inotersen) is an antisense oligonucleotide (ASO) that causes degradation of mutant and wild-type TTR mRNA through binding to the TTR mRNA, which results in a reduction of serum TTR protein and TTR protein deposits in tissues. By interfering with the RNA that carries damaged sequences for hATTR amyloidosis, it prevents the formation of amyloid fibrils. Because of the risks of serious bleeding caused by severe thrombocytopenia and because of glomerulonephritis, both of which require frequent monitoring, Patients using Tegsedi (inotersen) will need frequent testing for platelet counts and kidney function before, during and after treatment. Tegsedi (inotersen) is available only through a restricted distribution program under a Risk Evaluation and Mitigation Strategy (REMS) called the TEGSEDI REMS Program.

PHARMACY COVERAGE GUIDELINES  
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 2/21/2019  
LAST REVIEW DATE: 8/19/2021  
LAST CRITERIA REVISION DATE: 8/19/2021  
ARCHIVE DATE:

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

Vyndamax (tafamidis) or Vyndaquel (tafamidis meglumine) is 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.

The active ingredient of both Vyndamax (tafamidis) and Vyndaquel (tafamidis meglumine) is tafamidis. Tafamidis is a TTR stabilizer that selectively binds to TTR at the thyroxine binding sites and stabilizes the tetramer of the TTR transport protein, slowing dissociation into monomers that is the rate-limiting step in the amyloidogenic process. Tafamidis stabilizes both wild-type TTR tetramers and the tetramers of 14 TTR variants when tested clinically as well as 25 TTR variants tested ex vivo. Tafamidis is an analog of diflunisal that does not have anti-inflammatory properties.

Transthyretin amyloidosis is a slowly progressive condition characterized by the buildup of abnormal deposits of amyloid (amyloidosis) in organs and tissues. Protein deposition most frequently occurs in the peripheral nervous system resulting in a loss of sensation in the extremities (peripheral neuropathy). The autonomic nervous system may also be affected by amyloidosis. In some cases, the brain and spinal cord are affected. Other areas of amyloidosis include the heart, kidneys, eyes, and gastrointestinal tract. The age at which symptoms begin to develop varies widely among individuals with this condition and is typically between ages 20 and 70. The condition is inherited in an autosomal dominant pattern. The disease is caused by a mutation of the *TTR* gene located on chromosome 18 where valine is replaced by methionine at position 30 (TTR V30M or Val30Met). There are more than 130 mutations described.

The forms of transthyretin amyloidosis are distinguished by symptoms and the body system affected. The neuropathic form primarily affects the peripheral and autonomic nervous systems, resulting in peripheral neuropathy and difficulty controlling bodily functions. Impairments in bodily functions can include sexual impotence, diarrhea, constipation, problems with urination, and orthostatic hypotension. Some experience heart and kidney problems as well. Various eye problems may occur, such as cloudiness of the clear gel that fills the eyeball (vitreous opacity), dry eyes, glaucoma, or pupils with an irregular or "scalloped" appearance. Some also develop carpal tunnel syndrome, characterized by numbness, tingling, and weakness in the hands and fingers.

The leptomeningeal form primarily affects the central nervous system. In this form, amyloidosis occurs in the leptomeninges. Protein buildup can cause strokes and hemorrhage, hydrocephalus, ataxia, muscle stiffness and weakness (spastic paralysis), seizures, and loss of intellectual function. Eye problems similar to those in the neuropathic form may also occur.

The cardiac form primarily affects the heart which can lead to arrhythmias, cardiomegaly, or orthostatic hypertension. These abnormalities can lead to progressive heart failure and death. Occasionally, people with the cardiac form of transthyretin amyloidosis have mild peripheral neuropathy. The cardiac form can be hereditary or non-hereditary. The non-hereditary form is caused by aggregation of the wild-type transthyretin protein and is also known as Senile Systemic Amyloidosis.

Various stages of disease have been described. Patients with stage 0 disease are usually asymptomatic but have both a variant form of the TTR gene and evidence of amyloid deposits. Patients with stage I (mild) disease are still ambulatory, patients with stage II (moderate) disease are ambulatory but require assistance, and patients with stage III (severe) disease are bedridden or wheelchair bound.

PHARMACY COVERAGE GUIDELINES  
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 2/21/2019  
LAST REVIEW DATE: 8/19/2021  
LAST CRITERIA REVISION DATE: 8/19/2021  
ARCHIVE DATE:

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

Tissue biopsy should be used to confirm the diagnosis in all cases of amyloidosis, although the diagnosis of amyloidosis may be suspected on the basis of history and clinical manifestations. Tissue biopsy is done using Congo red staining and an immune histochemical study with anti-TTR antibodies. Genetic testing is needed to document the pathogenic mutation. If it is normal, TTR-FAP is excluded. Current techniques for performing sequence analysis of *TTR*, the only gene known to be associated with TTR amyloidosis, detect >99% of disease-causing mutations.

Orthotopic liver transplantation (OLT), which removes the main production site of the amyloidogenic protein, has historically been the standard of care for hereditary TTR amyloidosis. OLT is not effective in the non-neuropathic forms of familial TTR amyloidosis (i.e., cardiac amyloidosis, leptomeningeal amyloidosis, and familial oculoleptomeningeal amyloidosis [FOLMA]).

There are two other modes of treatment. The first is to reduce or halt the amount of mutant transthyretin that is synthesized through gene silencing by the liver. This approach employs use of small interfering RNA (patisiran) and antisense oligonucleotides (inotersen). The other approach is to stabilize mutant tetramers of transthyretin to prevent amyloidogenic monomers. Tetramer stabilizers include diflunisal and tafamidis. Currently under investigation is use of agent(s) to degrade amyloid fibrils that have already been deposited in tissues.

About 3,000 Americans are believed to have polyneuropathy caused by hATTR, which results from abnormally bent and folded proteins produced by mutated RNA. The amyloid fibrils (unusable proteins) deposit in nerves, where they produce pain in the arms, feet, hands and legs. Because they also accumulate in organ tissue, they can enlarge the heart and damage other organs.

---

**Definitions:**

**Risk Evaluation and Mitigation Strategy (REMS) Program**

Use of Tegsedi (inotersen) is subject to a Risk Evaluation and Mitigation Strategies (REMS) program that requires provider, patient, and dispensing pharmacy be enrolled into the program. Only providers and Pharmacies enrolled into the REMS may prescribe and dispense the drug, respectively, to individuals who are also in the program. A REMS program attempts to manage known or potentially serious risks associated with a drug product and is required by the Food and Drug Administration (FDA) for some drugs to ensure that the benefits of a drug outweigh its risks.

Because of the risks of serious bleeding caused by severe thrombocytopenia and because of glomerulonephritis, both of which require frequent monitoring, Tegsedi (inotersen) is available through a restricted REMS program

Requirements of the Tegsedi (inotersen) REMS Program include the following:

- Prescribers must be certified within the program by enrolling and completing training
- Patients must enroll in the program and comply with ongoing monitoring requirements
- Pharmacies must be certified with the program and must only dispense to patients who are authorized to receive TEGSEDI.

**Other names for polyneuropathy amyloidosis:**

Familial amyloid polyneuropathy type I (Portuguese-Swedish-Japanese type)  
Familial amyloid polyneuropathy type II (Indiana/Swiss or Maryland/German type)



PHARMACY COVERAGE GUIDELINES  
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 2/21/2019  
LAST REVIEW DATE: 8/19/2021  
LAST CRITERIA REVISION DATE: 8/19/2021  
ARCHIVE DATE:

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

Familial TTR amyloidosis  
Amyloid transthyretin polyneuropathy (ATTR-PN)  
Familial amyloidotic polyneuropathy (FAP)

**Staging:**

Clinical staging of TTR-FAP	
Stage 0	No symptoms
Stage I	Unimpaired ambulation; mostly mild sensory, motor, and autonomic neuropathy in the lower limbs
Stage II	Assistance with ambulation required; mostly moderate impairment progression to the lower limbs, upper limbs, and trunk
Stage III	Wheelchair-bound or bedridden; severe sensory, motor, and autonomic involvement of all limbs

**Polyneuropathy Disability Staging:**

Polyneuropathy Disability Stage	
Stage	Description
0	No symptoms
I	Sensory disturbances but preserved walking capability
II	Impaired walking capacity but ability to walk without a stick or crutches
IIIA	Walking with the help of one stick or crutch
IIIB	Walking with the help of two sticks or crutches
IV	Confined to a wheelchair or bedridden

**Vitamin A:**

Vitamin A (retinol, retinoic acid) is a nutrient important to vision, growth, cell division, reproduction and immunity. Vitamin A is found in many foods, such as spinach, dairy products and liver. Other sources are foods rich in beta-carotene, such as green leafy vegetables, carrots and cantaloupe. Your body converts beta-carotene into vitamin A. The recommended daily amount of vitamin A is 900 micrograms (mcg) for adult men and 700 mcg for adult women.

<https://ods.od.nih.gov/factsheets/VitaminA-Consumer/>

Currently, vitamin A is listed on food and supplement labels in international units (IUs) even though nutrition scientists rarely use this measure. Conversion rates between mcg retinol activity equivalents (RAE) and IU are as follows:

- 1 IU retinol = 0.3 mcg RAE
- 1 IU beta-carotene from dietary supplements = 0.15 mcg RAE
- 1 IU beta-carotene from food = 0.05 mcg RAE
- 1 IU alpha-carotene or beta-cryptoxanthin = 0.025 mcg RAE

**Resources:**

Tegsedi (inotersen) product information, revised by Akcea Therapeutics, Inc. 09-2020. Available at DailyMed <http://dailymed.nlm.nih.gov>. Accessed on July 10, 2021.



An Independent Licensee of the Blue Cross Blue Shield Association

PHARMACY COVERAGE GUIDELINES  
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 2/21/2019  
LAST REVIEW DATE: 8/19/2021  
LAST CRITERIA REVISION DATE: 8/19/2021  
ARCHIVE DATE:

---

**TEGSEDI™ (inotersen) subcutaneous injection**  
**VYNDAMAX™ (tafamidis) oral**  
**VYNDAQEL® (tafamidis meglumine) oral**

---

Vyndaqel (tafamidis meglumine) and Vyndamax (tafamidis) product information, revised by Pfizer Laboratories Div Pfizer Inc 06-2021. Available at DailyMed <http://dailymed.nlm.nih.gov>. Accessed on July 10, 2021.

Gorevic PD. Overview of amyloidosis. In: UpToDate, Lachmann HJ, Romain PL (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at <http://uptodate.com>. Accessed on July 10, 2021.

Fontana M. Amyloid cardiomyopathy: Treatment and prognosis. In: UpToDate, Rajkumar SV, McKenna WJ, Yeon SB (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at <http://uptodate.com>. Accessed on July 10, 2021.

Castano A, Helmke S, Alvarez J, et al.: Diflunisal for ATTR cardiac amyloidosis. Congest Heart Fail. 2012;18 (6): 315–319. Accessed January 03, 2020. Re-reviewed on July 10, 2021.

Berk JL, Suhr OB, Obici L, et al.: Repurposing Diflunisal for Familial Amyloid Polyneuropathy A Randomized Clinical Trial. JAMA 2013; 310 (24):2658-2667. Accessed November 09,2018. Re-reviewed on July 10, 2021.

---