

## CARDIAC AMYLOIDOSIS

2023 ACC Expert Consensus Decision Pathway on  
Comprehensive Multidisciplinary Care for the Patient with Cardiac Amyloidosis

Since the original publication of this document, acoramidis and vutisiran have been approved by the FDA for treatment of ATTR cardiac amyloidosis.

**Amyloidosis is a disease characterized by deposition of abnormal protein in the body's tissues and organs.** It can manifest in many different signs and symptoms depending on where the deposits are located.

When these proteins build up in the heart, it is known as cardiac amyloidosis.<sup>1</sup> Due to the complexity of cardiac amyloidosis, diagnosis and treatment ideally involves a multidisciplinary team.

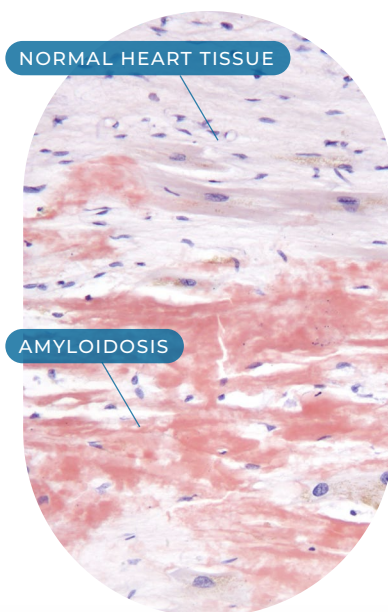


Figure A

**Amyloidosis** is the accumulation of abnormally folded proteins in tissues and organs such as the heart.



## OVERVIEW

### *Q: What is the purpose of this expert consensus decision pathway?*




The American College of Cardiology recently published an expert consensus decision pathway on comprehensive multidisciplinary care for patients with cardiac amyloidosis.<sup>1</sup> This decision pathway is designed to assist physicians in diagnosing and managing cardiac amyloidosis, emphasizing the importance of a multidisciplinary approach.

### *Q: What are the main types of cardiac amyloidosis?*

The two most common types of cardiac amyloidosis account for 90-95% of cases: amyloid light chain, or AL, and amyloid transthyretin, or ATTR.<sup>1</sup> Abnormal protein deposits in AL are caused by a protein known as monoclonal immunoglobulin light chain, which is produced by certain blood cells. Abnormal protein deposits in ATTR are caused by a protein known as transthyretin that is important for the function of transporting vitamin A and the thyroid gland.

### *Q: What are the main steps of the new expert consensus decision pathway?*

The decision pathway involves 3 steps.

- 
**1 Seek accurate diagnosis** and implement a cardiac treatment strategy.
- 
**2 Identify opportunities for collaboration** with other specialists to monitor the disease.
- 
**3 Improve access** to optimal care for amyloidosis.

## DIAGNOSIS

### *Q: What challenges do providers face in diagnosing a patient with cardiac amyloidosis?*

Several factors make the diagnosis of cardiac amyloidosis particularly difficult. The signs and symptoms of cardiac amyloidosis overlap with those of more common diseases. Additionally, patients with cardiac amyloidosis often experience multiple problems in other organs and systems that may seem unrelated. These can include kidney dysfunction, musculoskeletal conditions and neurological problems.



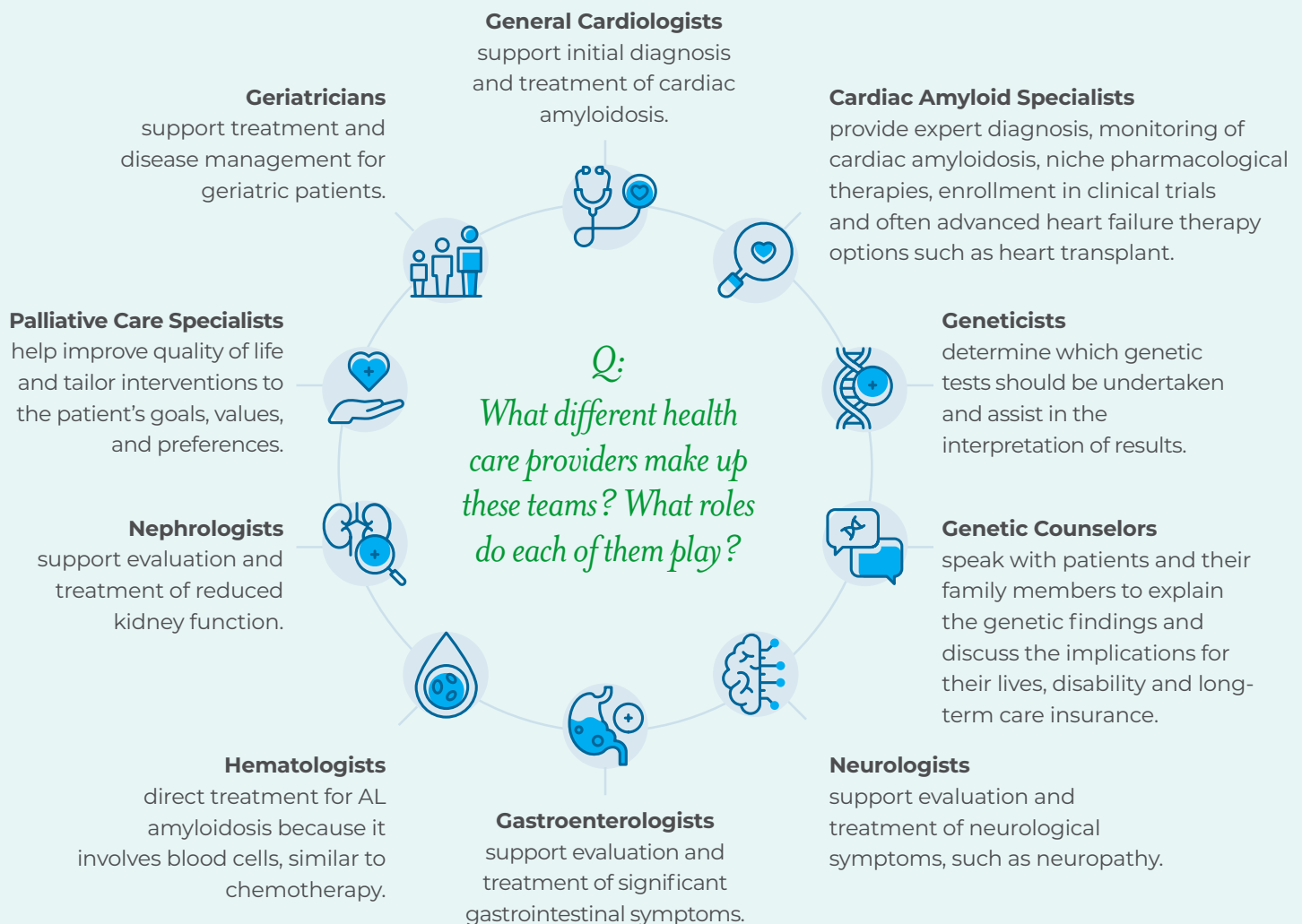
## HEALTH CARE PROVIDERS

*Q: Why is a multidisciplinary care team necessary?*

A multidisciplinary team is necessary because diagnosis and treatment of cardiac amyloidosis can be complicated. Many different organ systems are often involved, and long-term disease management may be necessary.

*Q: How does the expert consensus decision pathway suggest care teams collaborate to effectively care for the patient?*

Given that multiple specialists are often involved in treating patients with amyloidosis, it is important to identify a primary clinician to coordinate care. For patients with ATTR amyloidosis, this is typically a cardiologist. For patients with AL amyloidosis, this is typically a hematologist.



## TREATMENT & CARE MANAGEMENT

### *Q: What are the suggested treatments for cardiac amyloidosis?*

Treatment for cardiac amyloidosis involves addressing heart failure and arrhythmias, as well as initiating disease-modifying therapies that differ depending on the type of cardiac amyloidosis.

### TREATMENT OF HEART FAILURE AND ARRHYTHMIAS

#### Heart Failure



Loop diuretics



Mineralocorticoid receptor antagonists



Heart transplant



Palliative care

#### Arrhythmias



Pacemaker



Blood thinners



Implantable cardioverter-defibrillator therapies

### DISEASE-MODIFYING THERAPIES

#### ATTR cardiac amyloidosis



##### Tafamidis

- Stabilizes transthyretin protein and reduces abnormal accumulation in tissue
- Reduced death and hospitalization in clinical trials<sup>2</sup>
- Silencer therapies which reduce production of abnormal protein

#### AL cardiac amyloidosis

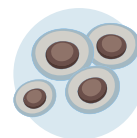


##### Chemotherapies and immunotherapies



##### Daratumumab

- When added to a regimen of other medications, can reduce deterioration of major organs such as the heart<sup>3</sup>



##### Stem cell therapy

## UNMET NEEDS

### *Q: What are the barriers to care for patients with cardiac amyloidosis?*

High medication cost is one of the main barriers to care for patients with cardiac amyloidosis. Most health plans cover medications approved for the treatment of cardiac amyloidosis. But patients may face prior authorization requirements, insurance approval delays and high out-of-pocket costs.

Another major barrier to care for patients with cardiac amyloidosis is limited access to specialists. Amyloid specialists are important in interpreting questionable and abnormal test results and helping direct care. Patients in rural areas and those unable to travel for medical care lack access to these specialists.

### *Q: What is the financial burden of care for cardiac amyloidosis?*

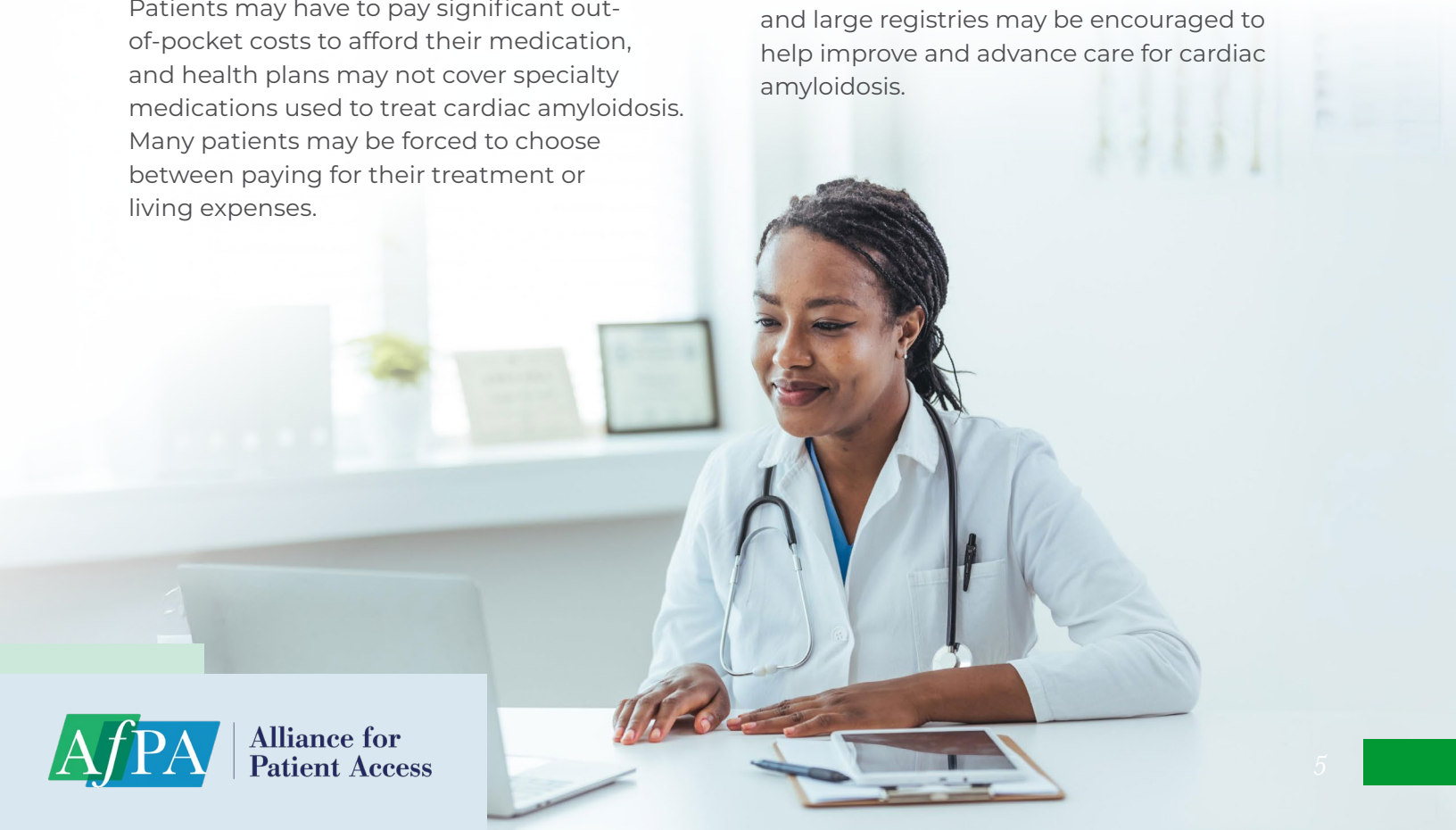
The financial burden of care for cardiac amyloidosis in the United States is immense. Patients may have to pay significant out-of-pocket costs to afford their medication, and health plans may not cover specialty medications used to treat cardiac amyloidosis. Many patients may be forced to choose between paying for their treatment or living expenses.

### *Q: How can telehealth help patients bridge potential gaps?*

Telehealth can enable access to specialists for patients who live in rural areas or are unable to travel to large medical centers for care. In such cases, amyloid specialists can partner with local clinicians via telehealth to discuss inconclusive testing and help determine subsequent tests and therapies.

### *Q: How can providers and policymakers work together to overcome unmet needs?*

For telehealth to be effectively integrated into health care, insurers must continue to allow physicians to bill telehealth visits as medical visits. State medical licensing rules should be revised to permit physicians to practice across state lines, allowing patients in rural areas to access care via telehealth. The creation of an online database of amyloidosis specialists may help increase referrals and improve specialty care. Finally, participation in clinical trials and large registries may be encouraged to help improve and advance care for cardiac amyloidosis.





This document was reviewed by cardiologist **Dharmesh Patel, MD, FACC, FASPC.**

## RESOURCES

1. Kittleson MM, Ruberg FL, Ambardekar AV, Brannagan TH, Cheng RK, Clarke JO, et al. 2023 ACC Expert Consensus Decision Pathway on Comprehensive Multidisciplinary Care for the Patient With Cardiac Amyloidosis: A Report of the American College of Cardiology Solution Set Oversight Committee. *J Am Coll Cardiol.* 2023;81(11):1076-126.
2. Maurer MS, Schwartz JH, Gundapaneni B, Elliott PM, Merlini G, Waddington-Cruz M, et al. Tafamidis Treatment for Patients with Transthyretin Amyloid Cardiomyopathy. *N Engl J Med.* 2018;379(11):1007-16.
3. Kastiris E, Palladini G, Minnema MC, Wechalekar AD, Jaccard A, Lee HC, et al. Daratumumab-Based Treatment for Immunoglobulin Light-Chain Amyloidosis. *N Engl J Med.* 2021;385(1):46-58.
4. Kazi DS, Bellows BK, Baron SJ, Shen C, Cohen DJ, Spertus JA, et al. Cost-Effectiveness of Tafamidis Therapy for Transthyretin Amyloid Cardiomyopathy. *Circulation.* 2020;141(15):1214-24.

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