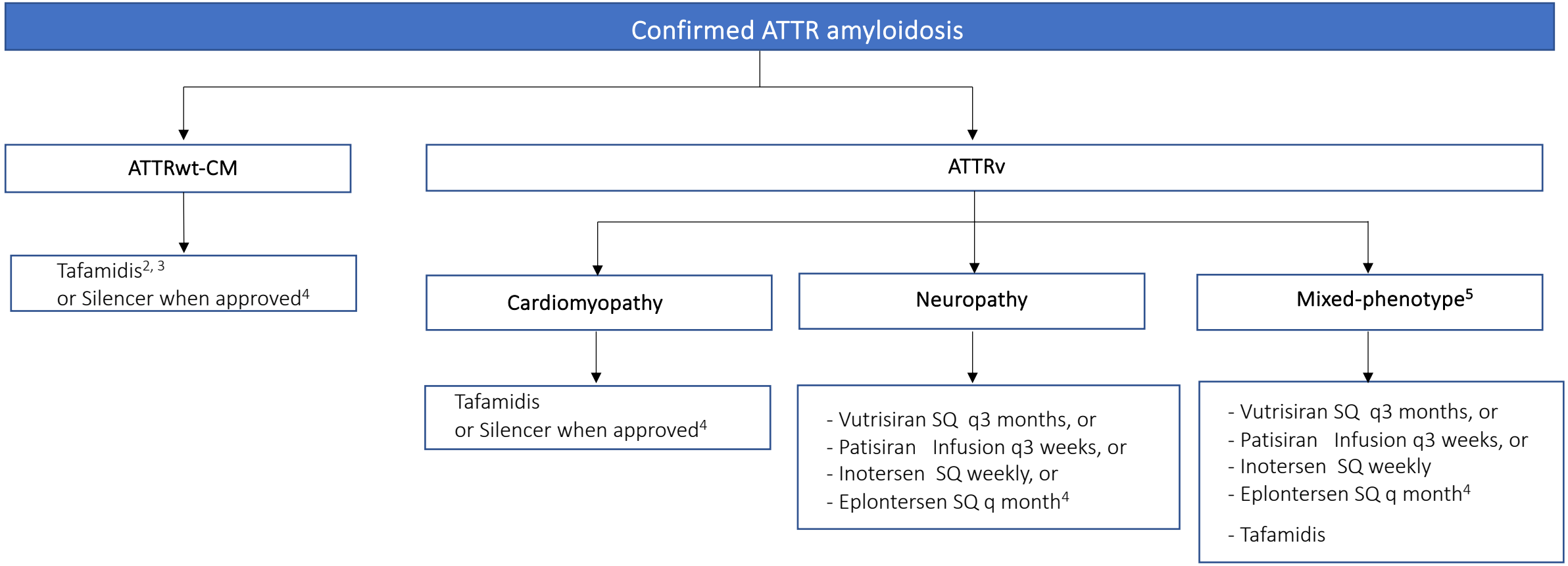
MAYO CLINIC

# Treatment for confirmed ATTR amyloidosis[1]




[1] Clinical trials, when possible, this algorithm lists commercially available options

[2] Benefits of Tafamidis in NYHA IV, end-stage renal disease are unclear.

[3] Off-label Diflunisal can be considered if the patient cannot tolerate or cannot access Tafamidis. Adding a proton pump inhibitor and monitoring renal function is recommended.

[4] Clinical scenarios, financial toxicity, and emerging data will guide the choice of stabilizer vs. silencer. The role of dual therapy is unknown, but single therapy is recommended due to cost and concerns regarding incremental benefit.

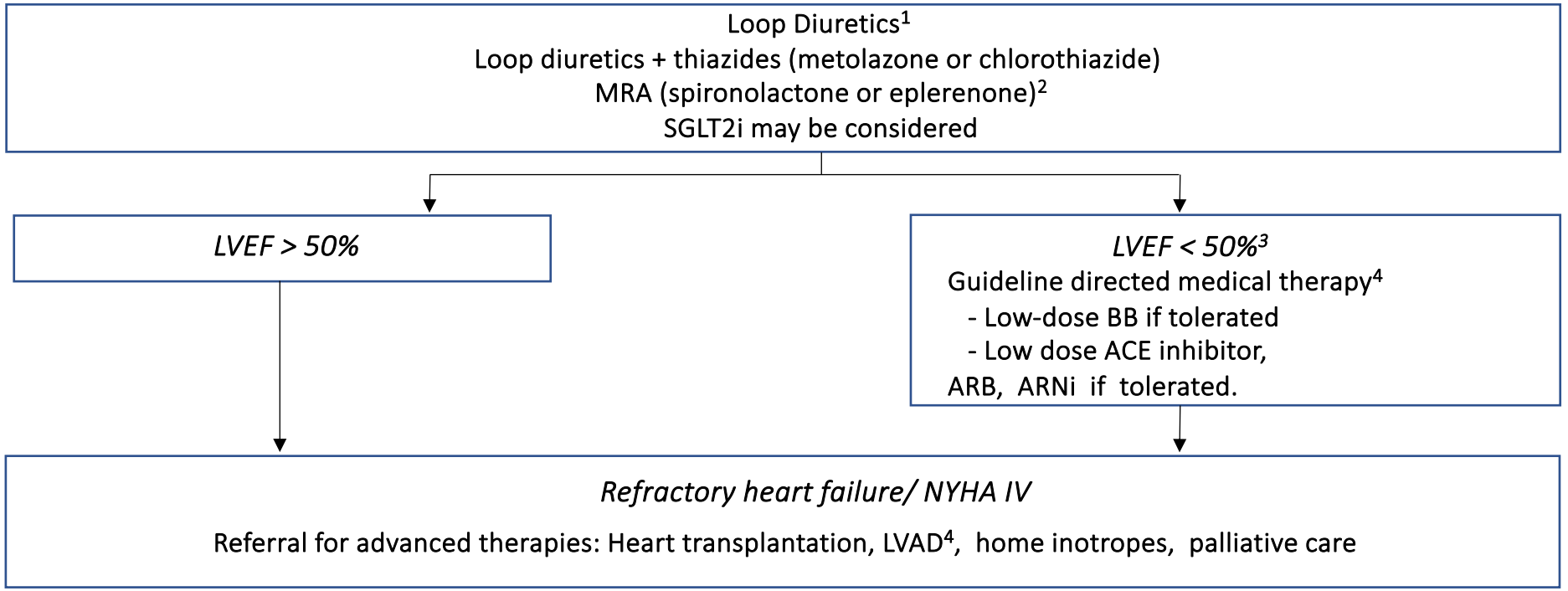
[5] Mixed phenotype is common; baseline cardiac and neurological evaluation is recommended for all. The choice of Rx is individualized.

ATTR, transthyretin amyloidosis; CM, cardiomyopathy; q, every; SQ, subcutaneous; v, variant; wt, wild type.

*Version 1.0 //last reviewed Dec 2023*

MAYO CLINIC

# Treatment of heart failure in ATTR amyloidosis[1]




[1] Loop diuretics with higher bioavailability (torsemide or bumetanide) are recommended.

[2] MRA are rarely discontinued and are associated with reduced mortality (*Ioanou, et al. Eur Heart J (2023) 44, 2893–2907*).

[3] Defibrillators can be considered in LVEF < 35%, but the benefit is unknown for primary prevention.

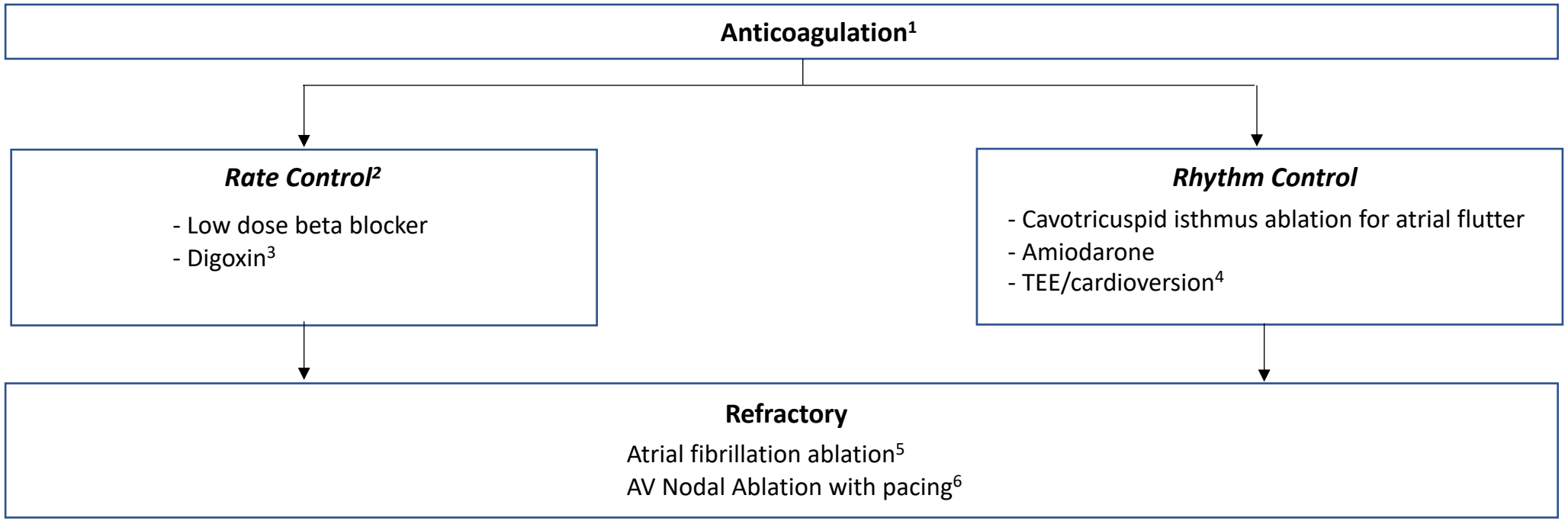
[4] Guideline directed medical therapy can worsen orthostatic hypotension, fatigue and dyspnea. BBs (if tolerated) are associated with reduced risk of mortality (*Ioanou, et al. Eur Heart J (2023) 44, 2893–2907*).

[5] LVAD could be considered in patients with remodeled ventricles with systolic dysfunction and dilated left ventricles.

ACE, angiotensin-converting enzyme inhibitor; ARB, angiotensin receptor blocker; ARNi, angiotensin receptor/neprilysin inhibitor; ATTR, transthyretin amyloidosis; BB, beta blocker; LVEF, left ventricular ejection fraction; MRA, mineralocorticoid receptor antagonists; SGLT2i, sodium/glucose cotransporter-2 inhibitors; NYHA, New York Heart Association.

MAYO CLINIC

# Treatment of Atrial flutter/fibrillation in ATTR amyloidosis




[1] Start anticoagulation independently of CHA2DS2-VASc score. Watchman devices are not recommended because of theoretical risk for thrombus formation.
[2] Non-dihydropyridine calcium channel blockers (diltiazem and verapamil) are contraindicated in amyloidosis.
[3] Low-dose digoxin with close monitoring. Goal trough level < 0.8 ng/l
[4] TEE is strongly suggested regardless of anticoagulation status. Cardioversion is usually unsuccessful.
[5] Atrial fibrillation ablation could be considered, with better response in early stages.
[6] Biventricular/his pacing is recommended.

ATTR, transthyretin amyloidosis; ; AV, atrio-ventricularTEE, transesophageal echocardiogram.

*Version 1.0 //last reviewed Dec 2023*

MAYO CLINIC

# Management of Asymptomatic TTR variant gene carriers

**Confirmed TTR gene carrier**

↓

**Assessment of Penetrance: start at least 10 years prior to the onset of symptoms in the first family member diagnosed**[1]

**Cardiac evaluation**
- History & physical examination
- 12 lead ECG
- NTproBNP, troponin, pre-albumin
- Echocardiogram
- Bone scintigraphy or CMR

**Neurological evaluation**
- EMG
- Autonomic reflex screen
- Neurology consult

↓

Repeat q 3-5 years in the absence of symptoms[2]
Repeat if symptoms arise
If tissue biopsies are done for any reason, consider stain for amyloid

[1] Age to screen remains uncertain. Family history and published variant-specific data may help. Testing may be tailored according to age and potentially related symptoms.
[2] Clinical annual evaluation could be considered

CMR, cardiac magnetic resonance; ECG, electrocardiogram; EMG, electromyography; q, every; TTR, transthyretin; .

MAYO CLINIC

# Asymptomatic patients with TTR deposits in other organs

**TTR deposits in other organs (e.g. carpal tunnel ligament, GI, etc.)**

↓

***Assessment of Penetrance***

**Cardiac evaluation**
- History & physical examination
- 12 lead ECG
- NTproBNP, troponin, prealbumin
- Echocardiogram
- Bone scintigraphy or CMR

**Neurological evaluation**
- EMG
- Autonomic reflex screen
- Neurology consult

↓

Repeat every 3-5 years in the absence of symptoms
Repeat if symptoms arise
If tissue biopsies are done for any reason, consider stain for amyloid

CMR, cardiac magnetic resonance; ECG, electrocardiogram; EMG, electromyography; TTR, transthyretin.