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HEALTH SCIENCES CENTER™

Amyloid Cardiomyopathy in West Texas

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Disclosures

- Nothing to disclose

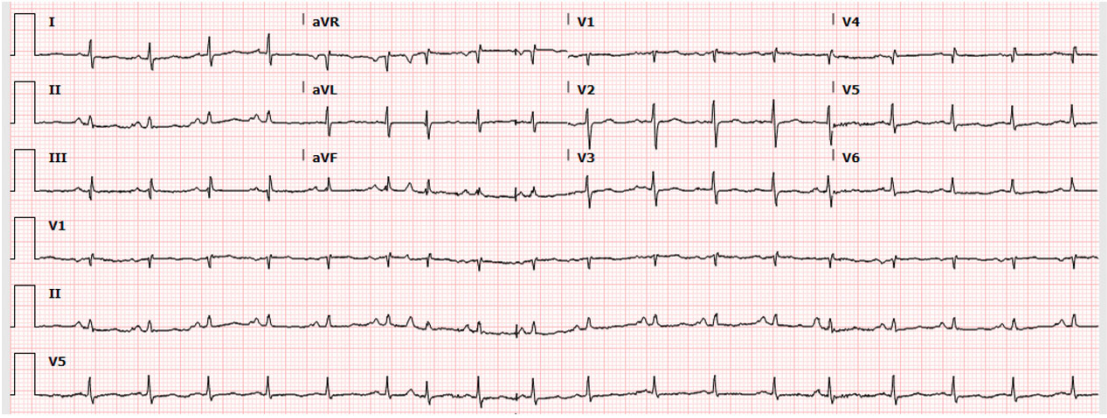
Objectives

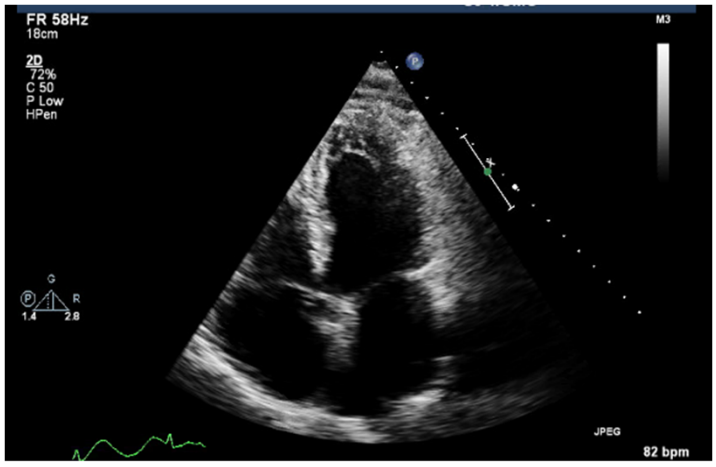
- Create awareness for amyloid cardiomyopathy
- Define amyloid cardiomyopathy subtypes
- Discuss treatment options for amyloid cardiomyopathy

Clinical Case

- 62-year-old AA man
  - Referred for chronic congestive heart failure symptoms, DOE, LE edema
  - LV EF 52%, LVIDd 5.0 cm; HFpEF; BMI 31
  - Medical Hx:
    - HTN
    - Atrial flutter s/p ablation (46y), AV block s/p PPM (59y), NSVT
    - ED
    - Bilateral carpal tunnel (45y)
    - Hx of retinal detachment
    - Recent prostate Ca Dx.
  - Social Hx: Previous runner, worked as a police officer
  - Family Hx: No premature heart disease.

Clinical Case





Clinical Case

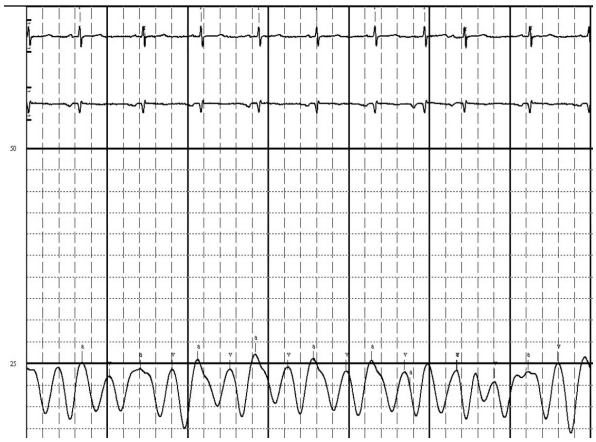
- Coronary angiography did not reveal obstructive CAD.
- Due to neuropathy with an unclear cause and his progressive heart failure symptoms genetic screening for hereditary ATTR was sent.

- Genetic testing:
  - Pathogenic mutation **p.V142I** in the TTR gene was identified.
  - Non invasive testing for cardiac involvement/cardiac amyloidosis included a PYP scan which was inconclusive.
  - He underwent a RHC with endomyocardial Bx.

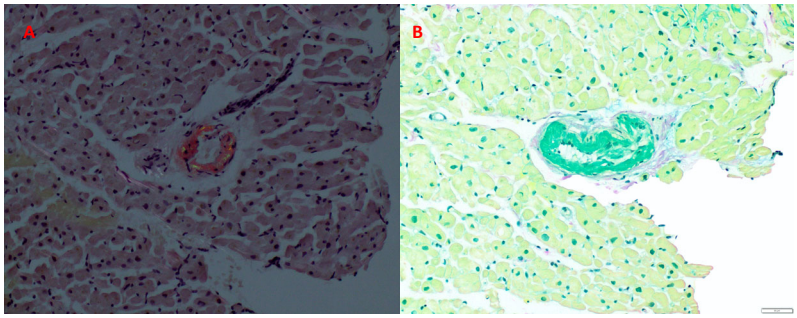
Hemodynamics

RA 22, RV 42/15, PA 42/25/32, PW 26,

TD CI 2.2



- Dx hereditary ATTR CM, confirmed by mass spectrometry



Juarez M, et al. J Prim Care Community Health. 2022. doi: 10.1177/21501319211062682.

Amyloid cardiomyopathy

- It is the result of misfolded protein in the heart, other systems affected are the nervous system.

- Two main subtypes:
  - Transthyretin amyloidosis (ATTR)
    - Hereditary (v - variant)
    - Wild type (wt - senile)
  - Light-chain amyloidosis (AL)

  - Other A Amyloidosis; secondary amyloidosis (AA)
    - Serum amyloid A protein; RA, Juvenile idiopathic arthritis, Ankylosing spondylitis, IBS, chronic infections, neoplasms.

Transthyretin (TTR) and Cardiomyopathy

- Produced in the liver
- Trans (transports) thy (thyroid hormone) retin (retinol).
- TTR also known as pre-albumin, migrates anodally to albumin on electrophoresis.
- Tetramer made of monomers that form a beta-pleated structure.
- The monomers become unstable, misfold and deposit in tissues forming amyloid fibrils.
- hATTR is inherited in an autosomal dominant fashion with age dependent incomplete penetrance.
- There are >100 variants.

Diagnosis

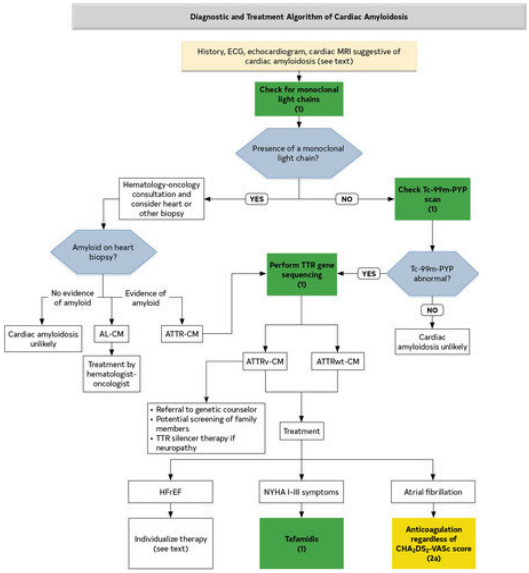
Recommendations for Diagnosis of Cardiac Amyloidosis

Referenced studies that support the recommendations are summarized in the Online Data Supplements.

COR	LOE	Recommendations
1	B-NR	1. Patients for whom there is a clinical suspicion for cardiac amyloidosis*[1–5] should have screening for serum and urine monoclonal light chains with serum and urine immunofixation electrophoresis and serum free light chains.[6]
1	B-NR	2. In patients with high clinical suspicion for cardiac amyloidosis, without evidence of serum or urine monoclonal light chains, bone scintigraphy should be performed to confirm the presence of transthyretin cardiac amyloidosis.[7]
1	B-NR	3. In patients for whom a diagnosis of transthyretin cardiac amyloidosis is made, genetic testing with *TTR* gene sequencing is recommended to differentiate hereditary variant from wild-type transthyretin cardiac amyloidosis.[8]

*LV wall thickness ≥14 mm in conjunction with fatigue, dyspnea, or edema, especially in the context of discordance between wall thickness on echocardiogram and QRS voltage on ECG, and in the context of aortic stenosis, HFpEF, carpal tunnel syndrome, spinal stenosis, and autonomic or sensory polyneuropathy.

Paul A. Heidenreich. Circulation. 2022 AHA/ACC/HFSA Guideline for the Management of Heart Failure: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines, Volume: 145, Issue: 18, Pages: e895-e1032, DOI: (10.1161/CIR.0000000000001063)



Paul A. Heidenreich. Circulation. 2022 AHA/ACC/HFSA Guideline for the Management of Heart Failure: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines, Volume: 145, Issue: 18, Pages: e895-e1032, DOI: (10.1161/CIR.0000000000001063)

Diagnosis

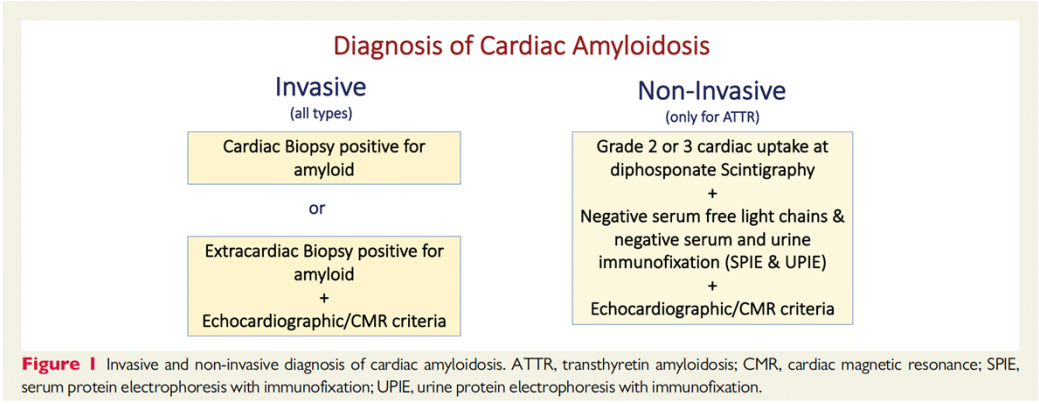


Figure 1 Invasive and non-invasive diagnosis of cardiac amyloidosis. ATTR, transthyretin amyloidosis; CMR, cardiac magnetic resonance; SPIE, serum protein electrophoresis with immunofixation; UPIE, urine protein electrophoresis with immunofixation.

Pablo Garcia-Pavia, et al. Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC Working Group on Myocardial and Pericardial Diseases, *European Heart Journal*, Volume 42, Issue 16, 21 April 2021, Pages 1554–1568

Red flag symptoms

Table 5
Cardiac and extracardiac amyloidosis red flags

Type	Red flag	Amyloidosis where it is most frequently found
Extracardiac		
Clinical	Polyneuropathy	ATTRv, AL, AA, AGel
	Dysautonomia	ATTR, AL
	Skin bruising	AL
	Skin discoloration	AApoAI
	Cutis laxa	AGel
	Macroglossia	AL
	Deafness	ATTRwt
	Bilateral carpal tunnel syndrome	ATTRv, ATTRwt
	Ruptured biceps tendon	ATTRwt
	Lumbar spinal stenosis	ATTRwt
	Vitreous deposits	ATTRv
	Corneal lattice dystrophy	AGel
	Family history	ATTRv, AApoAI, AApoAII
Laboratory	Renal insufficiency	AL, AA, AApoAI, AApoAII, AApoAIV, Aβ2M, AFib
	Proteinuria	AL, AA, AApoAI, AApoAII, AFib
Cardiac		
Clinical	Hypotension or normotensive if previous hypertensive	ATTR, AL
ECG	Pseudoinfarct pattern	All
	Low/decreased QRS voltage to degree of LV thickness	All
	AV conduction disease	All

Cardiac		
Clinical	Hypotension or normotensive if previous hypertensive	ATTR, AL
ECG	Pseudoinfarct pattern	All
	Low/decreased QRS voltage to degree of LV thickness	All
	AV conduction disease	All
Laboratory	Disproportionally elevated NT-proBNP to degree of HF	All
	Persisting elevated troponin levels	ATTR, AL
Echocardiogram	Granular sparkling of myocardium	All
	Increased right ventricular wall thickness	All
	Increased valve thickness	All
	Pericardial effusion	All
	Reduced longitudinal strain with apical sparing pattern	All
CMR	Subendocardial late gadolinium enhancement	All
	Elevated native T1 values	All
	Increased extracellular volume	All
	Abnormal gadolinium kinetics	All

Pablo Garcia-Pavia, et al. Diagnosis and treatment of cardiac amyloidosis: a position statement of the ESC Working Group on Myocardial and Pericardial Diseases, *European Heart Journal*, Volume 42, Issue 16, 21 April 2021, Pages 1554–1568

Transthyretin amyloid cardiomyopathy

Hereditary (hATTR; ATTRv)

- Associated to mutations in TTR gene.
- Initial symptoms include neuropathy.
- Cardiomyopathy presents at a later stage.

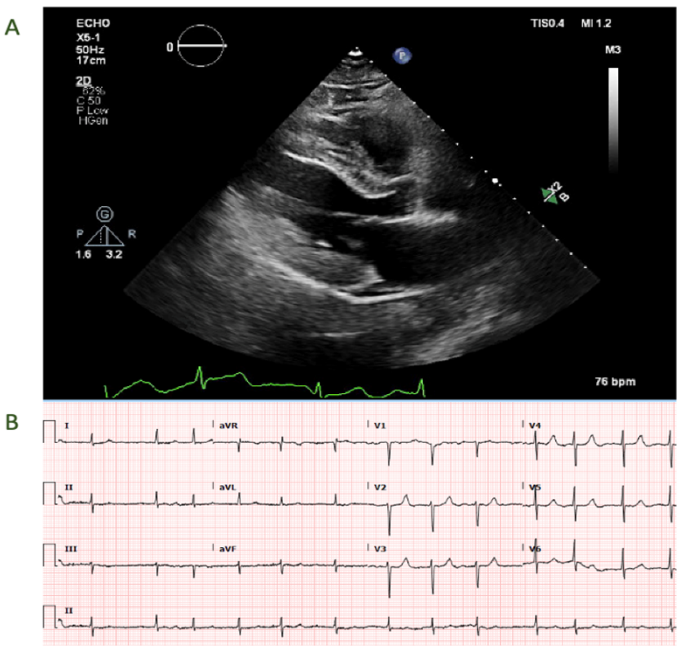
Wild type (ATTRwt)

- Presents at an older age.
- Also associated to neuropathy (spinal stenosis).
- Management includes stabilizing the TTR protein to avoid breakdown.

Clinical Case

- 85-year-old Caucasian male
  - He had been using furosemide for a long time on and off due to dyspnea and edema.
  - Found to have difficult to control atrial fibrillation
  - Past Medical Hx: PAF s/p ablation (81y), TIA, urinary retention/neurogenic bladder, No HTN
  - Familial Hx: No premature heart disease, no heart failure.
  - Social Hx: No recreational drug use, no tobacco

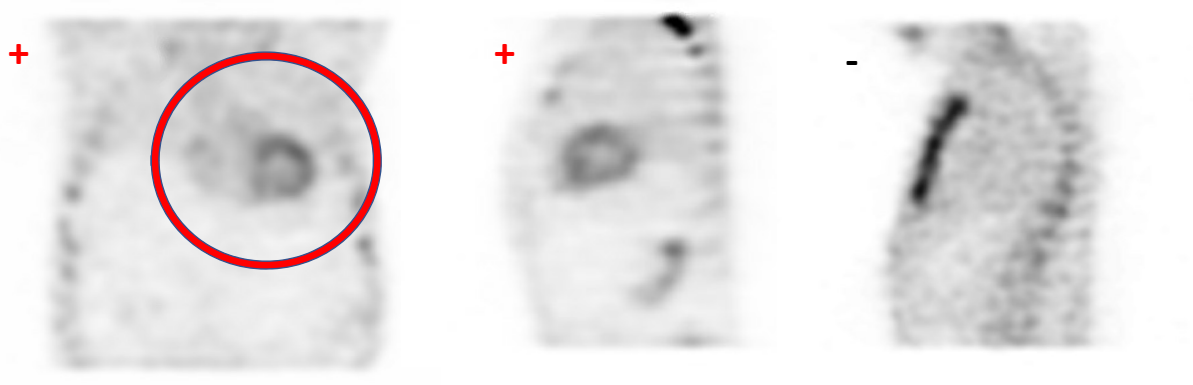
  - TTE newly diagnosed systolic heart failure, TTE LV EF 38%, LVH.
  - When in sinus rhythm his LV EF was normal, however diuretic requirements persisted.
  - No obstructive CAD on angiography.



Clinical Case

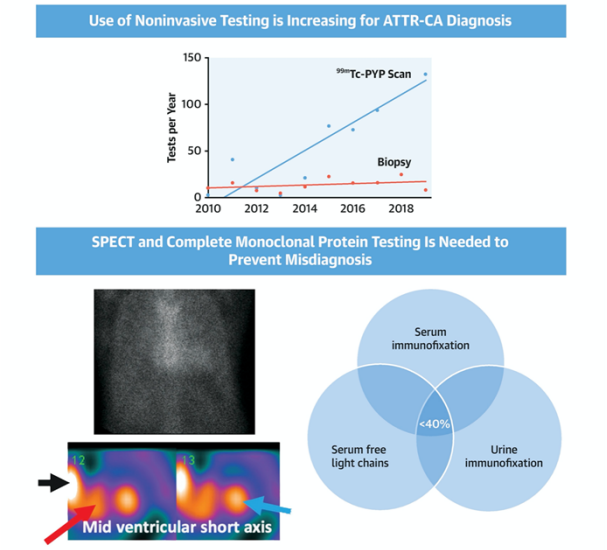
- Due to a discordant EKG and TTE with no Hx of hypertension it was decided to proceed with non invasive testing for amyloid cardiomyopathy (AL and ATTR).
  - No monoclonal proteins were identified (normal serum free light chain ratio, no monoclonal proteins identified on SPIE and UPIE)
  - PYP scan was abnormal.

Clinical Case



Genetic screening for hereditary types of ATTR is negative; He is diagnosed with wild type ATTR-CM.

CENTRAL ILLUSTRATION: Noninvasive Testing for ATTR-CA Is Increasing and the Use of SPECT and Monoclonal Protein Testing Is Crucial for Accurate Diagnosis



Poterucha, T.J. et al. J Am Coll Cardiol Img. 2021;14(6):1221-31.

Treatment/Management

- **Disease modifying therapy:**
  - Transthyretin silencers; Oligonucleotide (RNA) based; knock down TTR RNA that prevents hepatic production of the protein.
    - Patisiran
    - Vutisiran
    - Inotersen
  - Transthyretin stabilizers; prevent dissociation into monomers
    - Diflunisal
    - Tafamidis
  - Transthyretin disruptors; target tissue clearance
    - Doxyclyclone
    - Taurourodeoxycholic acid (TUDCA)
    - Epigallocatechin-3-gallate in green tea (EGCG)

JAMA Network™

From: **Repurposing Diflunisal for Familial Amyloid Polyneuropathy: A Randomized Clinical Trial**

JAMA. 2013;310(24):2658-2667. doi:10.1001/jama.2013.283815

Table 2. Longitudinal Intention-to-Treat Analyses of Primary (NIS+7) and Secondary Outcomes[a]

Outcomes	Mean (95% CI) Placebo Change From Baseline	Diflunisal Change From Baseline	Difference, Placebo–Diflunisal	P Value
NIS+7 score				
At 1 year	12.5 (8.6 to 16.4)	6.2 (2.8 to 9.6)	6.4 (1.2 to 11.6)	.02
At 2 years	26.3 (20.2 to 32.4)	8.2 (2.9 to 13.6)	18.0 (9.9 to 26.2)	<.001
NIS score				
At 1 year	10.1 (6.9 to 13.3)	4.1 (1.2 to 6.9)	6.0 (1.7 to 10.3)	.007
At 2 years	23.2 (17.8 to 28.5)	6.4 (1.6 to 11.2)	16.8 (9.6 to 24.0)	<.001
NIS-LL score				
At 1 year	6.0 (3.9 to 8.2)	3.2 (1.3 to 5.2)	2.8 (−0.1 to 5.7)	.06
At 2 years	12.1 (8.9 to 15.3)	3.8 (0.9 to 6.6)	8.3 (4.1 to 12.6)	<.001
Kumamoto score				
At 1 year	4.1 (2.1 to 6.2)	1.9 (0.1 to 3.7)	2.3 (−0.5 to 5)	.10
At 2 years	8.0 (5.8 to 10.3)	3.1 (1.1 to 5.1)	5.0 (1.9 to 8.0)	.002
Modified BMI[b]				
At 1 year	−38.5 (−74.9 to −2.1)	−18.7 (−51.6 to 14.1)	−19.8 (−68.8 to 29.2)	.43
At 2 years	−67.9 (−108.1 to −27.7)	−33.7 (−69.3 to 1.8)	−34.1 (−87.8 to 19.5)	.21
SF-36 physical component score				
At 1 year	−1.9 (−3.9 to 0.2)	0.7 (−1.1 to 2.5)	−2.6 (−5.3 to 0.1)	.06
At 2 years	−4.9 (−7.6 to −2.1)	1.2 (−1.2 to 3.7)	−6.1 (−9.8 to −2.5)	.001
SF-36 mental score				
At 1 year	0.8 (−2 to 3.6)	2.5 (0.0 to 5.1)	−1.7 (−5.5 to 2.1)	.37
At 2 years	−0.9 (−4.4 to 2.5)	3.5 (0.4 to 6.7)	−4.5 (−9.2 to 0.2)	.06

Abbreviations: BMI, body mass index; NIS, Neuropathy Impairment Score; NIS+7, NIS plus 7 nerve tests; NIS-LL, Neuropathy Impairment Score of the Lower Limbs; SF-36, 36-Item Short-Form Health Survey.

[a] Linear models for repeated measures of outcome data were used. Means were calculated for change from baseline to 12 and 24 months for primary and secondary outcome measures by treatment groups. *P* values address the differences between treatment groups in change over 12 and 24 months for each outcome measure. See Table 1 footnotes for explanation of score ranges.

[b] Modified BMI is the product of BMI (weight in kilograms divided by the square of height in meters) and serum albumin (g/L).

Table Title:
Longitudinal Intention-to-Treat Analyses of Primary (NIS+7) and Secondary Outcomes[a]Abbreviations: BMI, body mass index; NIS, Neuropathy Impairment Score; NIS+7, NIS plus 7 nerve tests; NIS-LL, Neuropathy Impairment Score of the Lower Limbs; SF-36, 36-Item Short-Form Health Survey.

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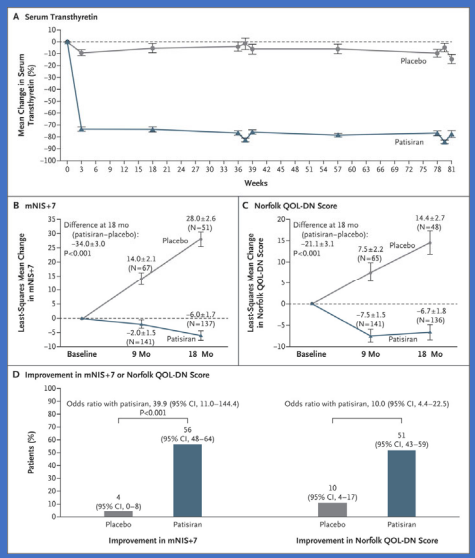
Comparisons of Changes between the Patisiran Group and the Placebo Group over Time.



D Adams et al. N Engl J Med 2018;379:11-21.

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Secondary and Exploratory End Points.

Table 2. Secondary and Exploratory End Points.

End Point	Placebo	Patisiran	Least-Squares Mean Difference (Patisiran – Placebo)	P Value
Secondary end points in the modified ITT population*				
No. of patients	77	148		
Neuropathy Impairment Score–weakness†				
Mean (±SD) baseline score	29.0±23.0	32.7±25.2		
Least-squares mean (±SE) change from baseline at 18 mo	17.9±2.0	0.1±1.3	−17.9±2.3	<0.001
Score on the Rasch-built Overall Disability Scale‡				
Mean (±SD) baseline score	29.8±10.8	29.7±11.5		
Least-squares mean (±SE) change from baseline at 18 mo	−8.9±0.9	0.0±0.6	9.0±1.0	<0.001
10-m walk test — m/sec§				
Mean (±SD) baseline value	0.79±0.32	0.80±0.40		
Least-squares mean (±SE) change from baseline at 18 mo	−0.24±0.04	0.08±0.02	0.31±0.04	<0.001
Modified BMI¶				
Mean (±SD) baseline value	989.9±214.2	969.7±210.5		
Least-squares mean (±SE) change from baseline at 18 mo	−119.4±14.5	−3.7±9.6	115.7±16.9	<0.001
Composite Autonomic Symptom Score 31‖				
Mean (±SD) baseline score	30.3±16.4	30.6±17.6		
[illegible]				
Exploratory end points in the cardiac subpopulation**				
No. of patients	36	90		
Left ventricular wall thickness — mm				
Mean (±SD) baseline value	16.4±2.1	16.8±2.6		
Least-squares mean (±SE) change from baseline at 18 mo	−0.1±0.3	−1.0±0.2	−0.9±0.4	0.02
Left ventricular longitudinal strain — %				
Mean (±SD) baseline value	−15.66±3.51	−15.13±3.41		
Least-squares mean (±SE) change from baseline at 18 mo	1.46±0.48	0.08±0.28	−1.37±0.56	0.02
NT-proBNP††				
Baseline value				
Geometric mean — pg/ml	711.1	726.9		
Coefficient of variation — %	190.8	220.3		
Ratio to baseline at 18 mo‡‡	1.97	0.89	0.45§§	<0.001

* The modified intention-to-treat (ITT) population included all the patients who underwent randomization and received at least one dose of patisiran or placebo.
† Scores on the weakness component of the Neuropathy Impairment Score range from 0 to 192, with higher scores indicating more impairment. The number of patients who were assessed at 18 months was 51 in the placebo group and 137 in the patisiran group.
‡ Scores on the Rasch-built Overall Disability Scale range from 0 to 48, with lower scores indicating more disability. The number of patients who were assessed at 18 months was 54 in the placebo group and 138 in the patisiran group.
§ A lower value indicates a slower gait speed. The number of patients who were assessed at 18 months was 55 in the placebo group and 138 in the patisiran group.
¶ The modified body-mass index (BMI) was the BMI (weight in kilograms divided by square of height in meters) × albumin level in grams per liter. The number of patients who were assessed at 18 months was 52 in the placebo group and 133 in the patisiran group.
‖ Values for the Composite Autonomic Symptom Score 31 range from 0 to 100, with higher scores indicating more autonomic symptoms. The number of patients who were assessed at 18 months was 53 in the placebo group and 136 in the patisiran group.
** The cardiac subpopulation included patients with a baseline left ventricular wall thickness of 13 mm or more in the absence of a history of aortic valve disease or hypertension.
†† N-terminal pro–brain natriuretic peptide (NT-proBNP) is a measure of cardiac stress that is an independent predictor of death in patients with transthyretin cardiac amyloidosis.
‡‡ Shown is the adjusted geometric mean ratio to baseline at month 18.
§§ Shown is the ratio of adjusted geometric mean ratio to baseline at month 18 between the two trial groups (patisiran:placebo).

D Adams et al. N Engl J Med 2018;379:11-21.

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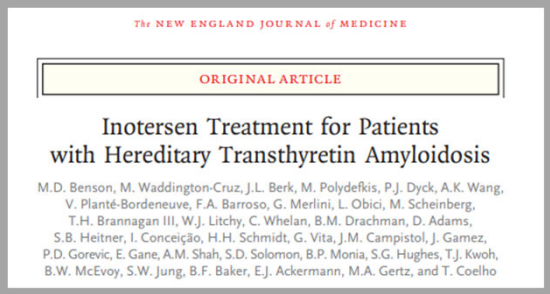
Change from Baseline in Primary End Points.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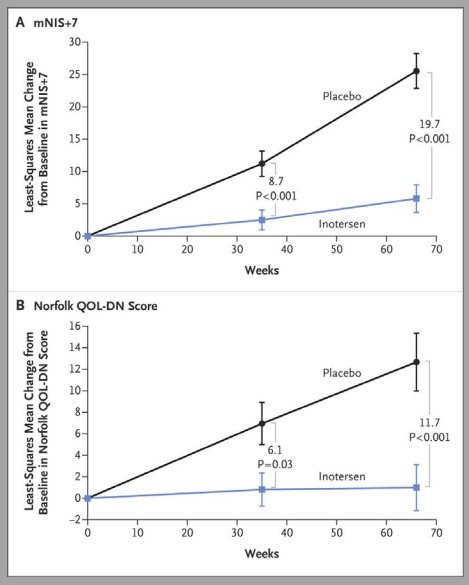
ORIGINAL ARTICLE

Inotersen Treatment for Patients with Hereditary Transthyretin Amyloidosis

M.D. Benson, M. Waddington-Cruz, J.L. Berk, M. Polydefkis, P.J. Dyck, A.K. Wang, V. Planté-Bordeneuve, F.A. Barroso, G. Merlini, L. Obici, M. Scheinberg, T.H. Brannagan III, W.J. Litchy, C. Whelan, B.M. Drachman, D. Adams, S.B. Heitner, I. Conceição, H.H. Schmidt, G. Vita, J.M. Campistol, J. Gamez, P.D. Gorevic, E. Gane, A.M. Shah, S.D. Solomon, B.P. Monia, S.G. Hughes, T.J. Kwoh, B.W. McEvoy, S.W. Jung, B.F. Baker, E.J. Ackermann, M.A. Gertz, and T. Coelho



MD Benson et al. N Engl J Med 2018;379:22-31.

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Primary Analysis and Components.

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ESTABLISHED IN 1812 SEPTEMBER 13, 2018 VOL. 379 NO. 11

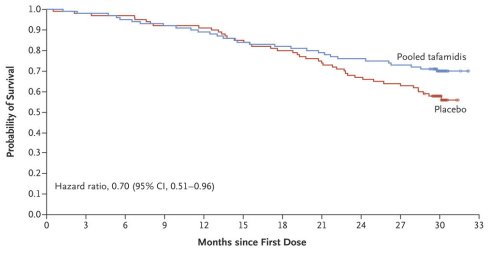
Tafamidis Treatment for Patients with Transthyretin Amyloid Cardiomyopathy

Mathew S. Maurer, M.D., Jeffrey H. Schwartz, Ph.D., Balarama Gundapaneni, M.S., Perry M. Elliott, M.D., Giampaolo Merlini, M.D., Ph.D., Marcia Waddington-Cruz, M.D., Arnt V. Kristen, M.D., Martha Grogan, M.D., Ronald Witteles, M.D., Thibaud Damy, M.D., Ph.D., Brian M. Drachman, M.D., Sanjiv J. Shah, M.D., Mazen Hanna, M.D., Daniel P. Judge, M.D., Alexandra I. Barsdorf, Ph.D., Peter Huber, R.Ph., Terrell A. Patterson, Ph.D., Steven Riley, Pharm.D., Ph.D., Jennifer Schumacher, Ph.D., Michelle Stewart, Ph.D., Marla B. Sultan, M.D., M.B.A., and Claudio Rapezzi, M.D., for the ATTR-ACT Study Investigators*

A Primary Analysis, with Finkelstein–Schoenfeld Method

	No. of Patients	P Value from Finkelstein–Schoenfeld Method	Win Ratio (95% CI)	Patients Alive at Mo 30 *no. (%)*	Average Cardiovascular-Related Hospitalizations during 30 Mo among Those Alive at Mo 30 *per patient per yr*
Pooled Tafamidis	264	<0.001	1.70 (1.26–2.29)	186 (70.5)	0.30
Placebo	177			101 (57.1)	0.46

B Analysis of All-Cause Mortality



No. at Risk (cumulative no. of events)

Pooled tafamidis	264 (0)	259 (5)	252 (12)	244 (20)	235 (29)	222 (42)	216 (48)	209 (55)	200 (64)	193 (71)	99 (78)	0 (78)
Placebo	177 (0)	173 (4)	171 (6)	163 (14)	161 (16)	150 (27)	141 (36)	131 (46)	118 (59)	113 (64)	51 (75)	0 (76)

C Frequency of Cardiovascular-Related Hospitalizations

	No. of Patients	No. of Patients with Cardiovascular-Related Hospitalizations *total no. (%)*	Cardiovascular-Related Hospitalizations *no. per yr*	Pooled Tafamidis vs. Placebo Treatment Difference *relative risk ratio (95% CI)*
Pooled Tafamidis	264	138 (52.3)	0.48	0.68 (0.56–0.81)
Placebo	177	107 (60.5)	0.70	

MS Maurer et al. N Engl J Med 2018;379:1007-1016.

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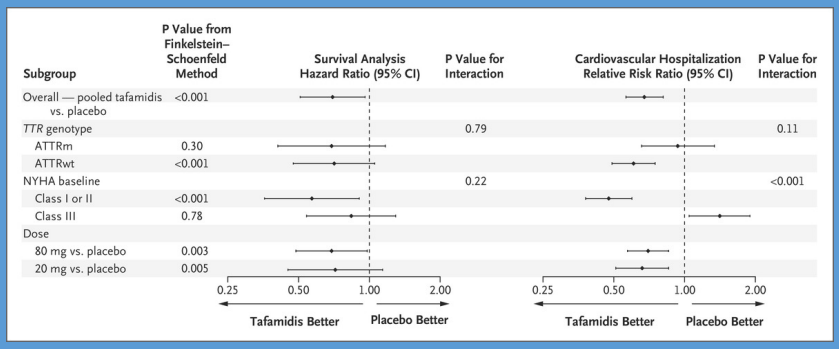
Overall and Subgroup Results as Calculated with the Use of the Finkelstein–Schoenfeld Method, All-Cause Mortality, and Cardiovascular-Related Hospitalizations.



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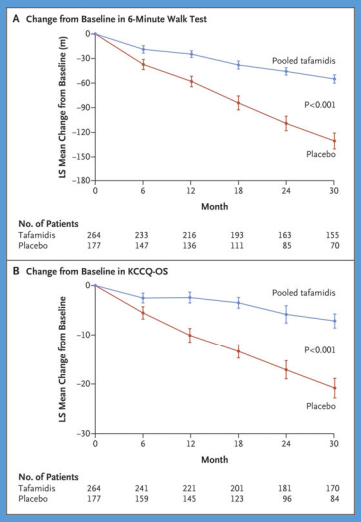
Key Secondary End Points.



MS Maurer et al. N Engl J Med 2018;379:1007-1016.

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Recommendations for Treatment of Cardiac Amyloidosis

Referenced studies that support the recommendations are summarized in the Online Data Supplements.

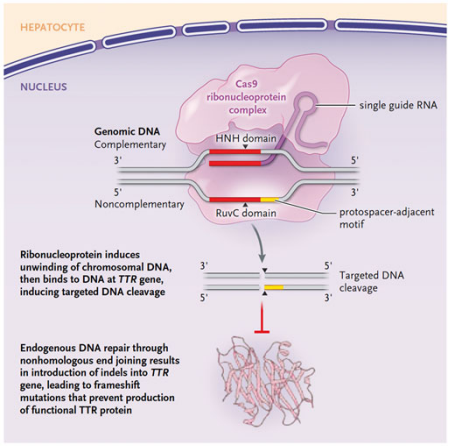
COR	LOE	Recommendations
1	B-R	1. In select patients with wild-type or variant transthyretin cardiac amyloidosis and NYHA class I to III HF symptoms, transthyretin tetramer stabilizer therapy (tafamidis) is indicated to reduce cardiovascular morbidity and mortality.[1]
Value Statement: Low Value (B-NR)		2. At 2020 list prices, tafamidis provides low economic value (>$180 000 per QALY gained) in patients with HF with wild-type or variant transthyretin cardiac amyloidosis.[2]
2a	C-LD	3. In patients with cardiac amyloidosis and AF, anticoagulation is reasonable to reduce the risk of stroke regardless of the CHA_2DS_2-VASc (congestive heart failure, hypertension, age ≥75 years, diabetes mellitus, stroke or transient ischemic attack [TIA], vascular disease, age 65 to 74 years, sex category) score.[3,4]

Paul A. Heidenreich. Circulation. 2022 AHA/ACC/HFSA Guideline for the Management of Heart Failure: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines, Volume: 145, Issue: 18, Pages: e895-e1032, DOI: (10.1161/CIR.0000000000001063)

Novel concepts

- Clustered regularly interspaced short palindromic repeats and associated Cas9 endonuclease (CRISPR-Cas9) system

- Gene editing therapy that reduces the production of transthyretin.



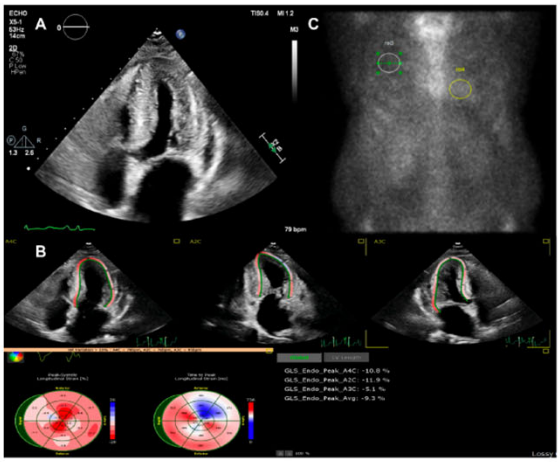
Gillmore JD et al. DOI: 10.1056/NEJMoa2107454

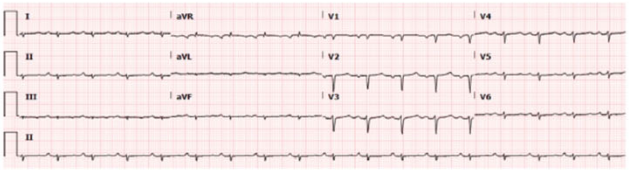
Clinical Case

- 58-year-old woman referred for lower extremity edema that was refractory to diuretics and dyspnea present for a year. Also recently diagnosed with proteinuria.
  - Medical Hx: Hypothyroidism.
  - Physical exam + Kussmaul sign, bilateral LE pitting edema.
  - Laboratory work significant for hypercholesterolemia, hypoalbuminemia and proteinuria 2.375 g/24hr.

  - Serum free light chain ratio (Kappa/Lambda) 0.15; Serum protein electrophoresis significant for elevated lambda monoclonal protein.

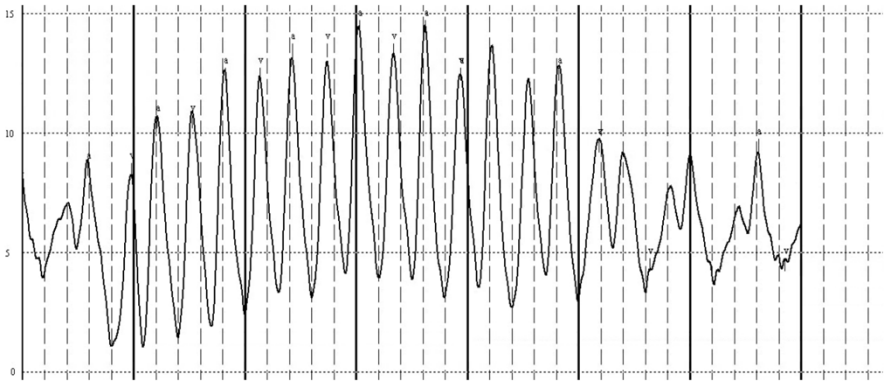
Clinical Case



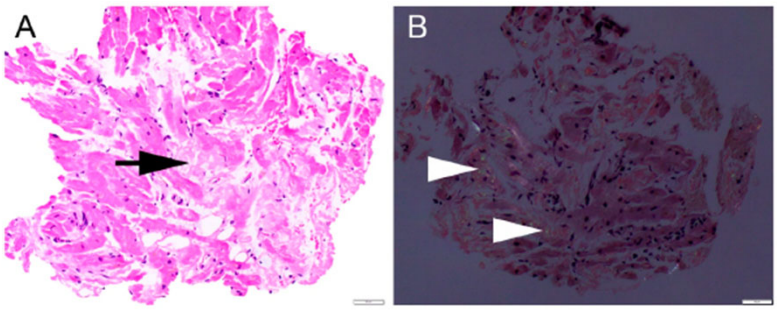


Parmar, K. Clinical Case Reports 2022. DOI: 10.22541/au.165484104.46215880/v1

Clinical Case: Hemodynamics



Clinical Case



Mass spectrometry revealed a profile consistent with AL (lambda) type amyloid deposition.

Light chain amyloid CM

- AL amyloidosis is associated to a hematologic disorder which includes plasma cell dyscrasias or B-Cell lymphoma.
- Referral hematology/oncology is necessary for ongoing care.
- The case presented underwent treatment with cyclophosphamide, bortezomib, dexamethasone + daratumumab. And is now being considered for bone marrow transplant.
- Additional considerations are screening for atrial fibrillation.

Take home points

- Amyloid cardiomyopathy is a differential diagnosis in patients with cardiovascular disease symptoms.
- There are different subtypes of amyloidosis.
- Identifying the subtype of amyloid cardiomyopathy can alter the management course of the patient.