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A CORUÑA 27-28 SEPTIEMBRE 2024

XI Meeting. State of the Art in
HEART FAILURE
CLINICAL PRACTICE AND ORGANIZATIONAL MODELS
Venue: Hotel Meliá MaríaPita, A Coruña
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A CORUÑA 27-28 SEPTEMBER 2024

PROGNOSTIC VALUE OF TRICUSPID ANNULAR PLANE SYSTOLIC EXCURSION/SYSTOLIC PULMONARY ARTERY PRESSURE RATIO IN CARDIAC AMYLOIDOSIS

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Humanitas Research Hospital, Milan, Italy

HFA meeting May 2023, Prague

Highlights session (Heart Failure Imaging)

Original article

Prognostic value of the tricuspid annular plane systolic excursion/systolic pulmonary artery pressure ratio in cardiac amyloidosis

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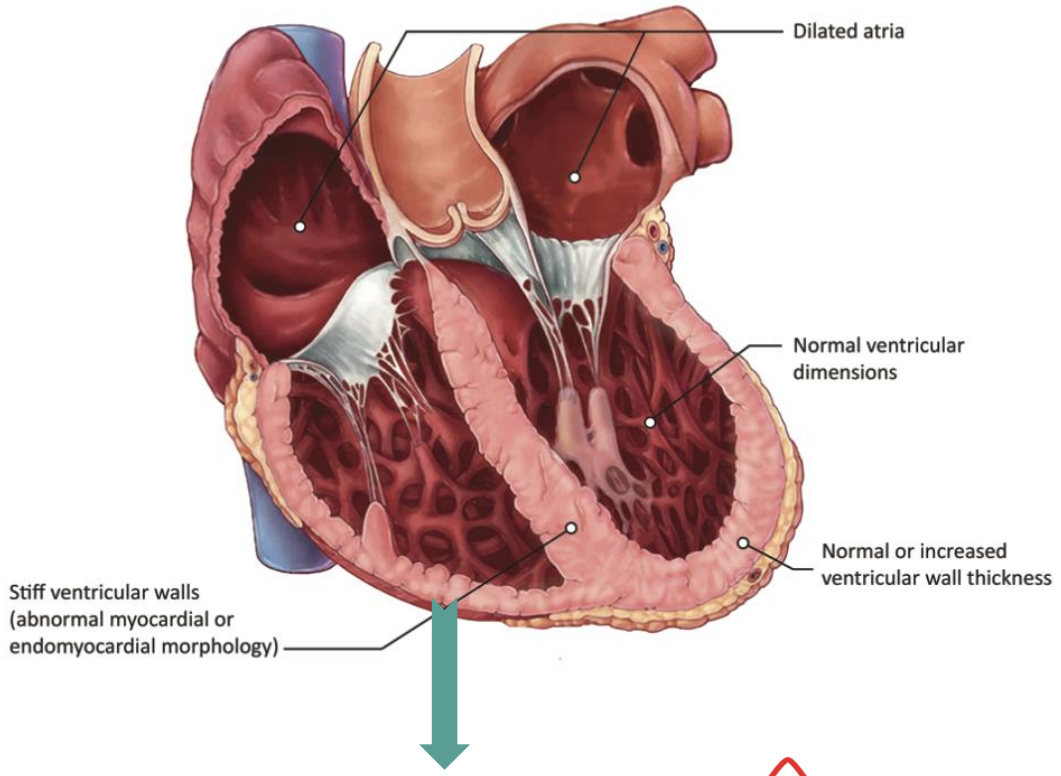
Maccallini M, et al. Rev Esp Cardiol. 2024;77:634-644. Epub 2024 Jan 29.

Background

The adequacy of **right ventricle adaptation** to increased **afterload** is a known determinant of the severity of symptoms and long-term outcomes in **heart failure or pulmonary hypertension**.

Restrictive Cardiomyopathy



Influence of the **Right Ventricle** on prognosis

Guazzi M, Dixon D, et al. JACC Cardiovasc Imaging. 2017 Oct; Ghio et al, Circ cardiovascular imaging, 2016

Background

Circulation: Cardiovascular Imaging

ORIGINAL ARTICLE

Validation of the Tricuspid Annular Plane Systolic Excursion/Systolic Pulmonary Artery Pressure Ratio for the Assessment of Right Ventricular-Arterial Coupling in Severe Pulmonary Hypertension

Different correlates but similar prognostic implications for right ventricular dysfunction in heart failure patients with reduced or preserved ejection fraction

Stefano Ghio, Marco Guazzi, Angela Beatrice Scardovi, Catherine Klersy, Francesco Clemenza, Erberto Carluccio, Pier Luigi Temporelli, Andrea Rossi, Pompilio Faggiano ... See all authors

First published: 17 November 2016 | https://doi.org/10.1002/ejhf.664 | Citations: 194

RV Contractile Function and its Coupling to Pulmonary Circulation in Heart Failure With Preserved Ejection Fraction

Stratification of Clinical Phenotypes and Outcomes

Marco Guazzi, MD, PhD,[a,b] Debra Dixon, BS,[c] Valentina Labate, MD,[a,b] Lauren Beussink-Nelson, RDCS, MHS Francesco Bandera, MD,[a,b] Michael J. Cuttica, MD,[d] Sanijv J. Shah, MD[c,e]

Tricuspid annular plane systolic excursion and pulmonary arterial systolic pressure relationship in heart failure: an index of right ventricular contractile function and prognosis

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Impact of Right Ventricle-Pulmonary Artery Coupling on Clinical Outcomes in the PARTNER 3 Trial

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Right Ventricular-Pulmonary Arterial Coupling and Afterload Reserve in Patients Undergoing Transcatheter Tricuspid Valve Repair

Michael I. Brener, MD,[a] Philipp Lurz, MD, PhD,[b] Jörg Hausleiter, MD,[c] Josep Rodés-Cabau, MD,[d] Neil Fam,

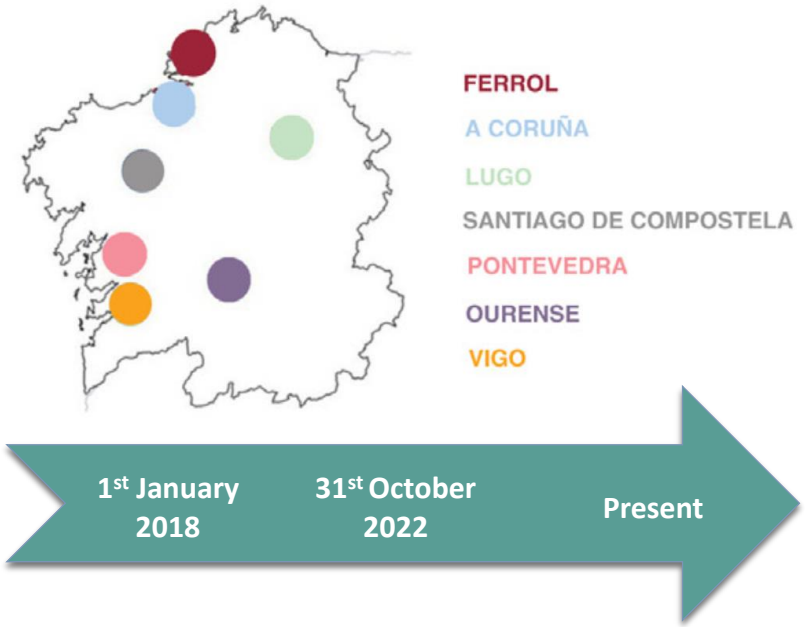
Guazzi M, Dixon D, et al. JACC Cardiovasc Imaging. 2017 Oct;10:1211–21.

Purpose

To describe distribution of the **TAPSE/SPAP ratio** and to evaluate its **prognostic implications** in a multi-institutional **cohort of patients with cardiac amyloidosis**.

AMIGAL (Registro de **AMI**loidosis cardiaca de **GAL**icia)

Prospective, observational registry of patients with A-CM (7 hospitals)



- **Inclusion criteria:** baseline echocardiographic data to calculate TAPSE/SPAP ratio
- **Follow-up**: from date of inclusion to 30th November 2022/death/heart transplantation (HTx)
- **Outcomes**: overall survival and survival free of heart failure hospitalization or HTx

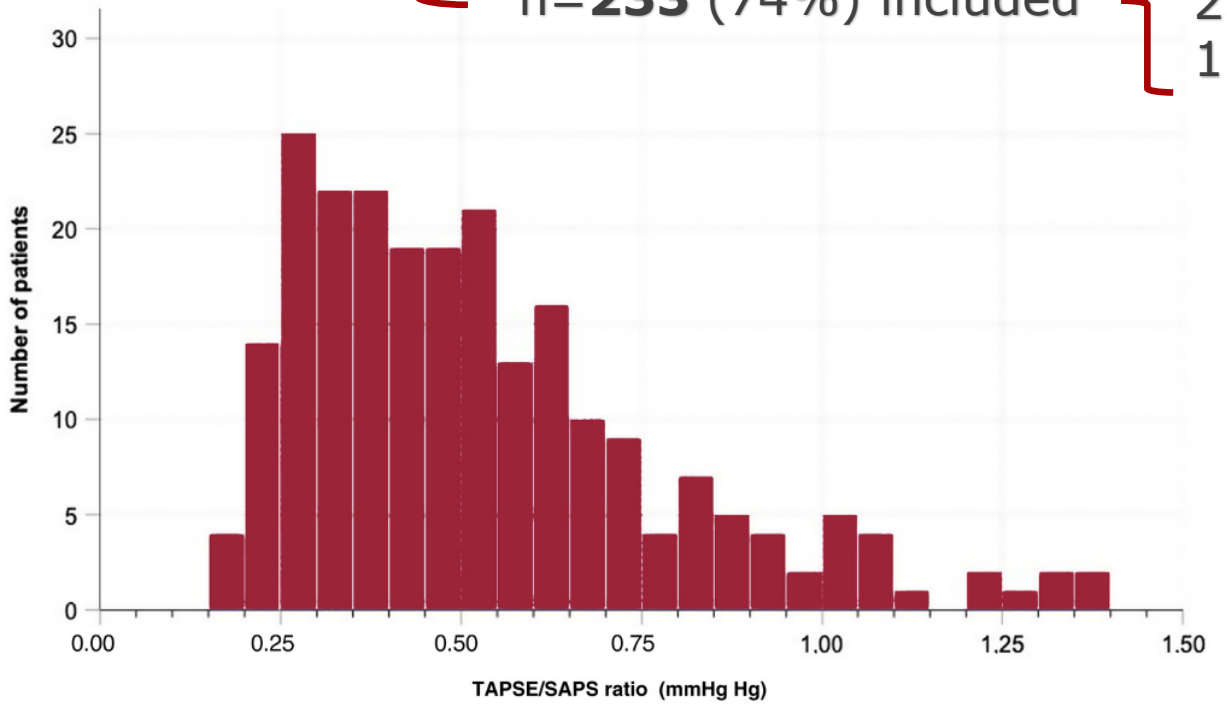
Results

AMIGAL study
n=315 patients
(31/10/22)

n=**82** excluded [72 (87.8%) for no/trace tricuspid regurgitation]

n=**233** (74%) included

209 (89.7%) **ATTR-CM (176-84.2% ATTRwt)**
23 (9.9%) **AL-CM**
1 (0.4%) ApoA-IV



Median: 0.48 (IQR 0.33-0.65)
Tertiles: <0.38, 0.38-0.58, >0.58

Figure 1. Distribution of baseline values of TAPSE/SPAP ratio in the study population.

Baseline characteristics

Variables	TAPSE/SPAP < 0.38 n = 77	TAPSE/SPAP 0.38 – 0.58 n = 78	TAPSE/SPAP > 0.58 n = 78	*P*
Medical history				
Age, y	79.9 ± 8.8	81.4 ± 6.4	79.8 ± 7.5	.340
Women	25 (32.5)	22 (28.2%)	16 (20.5%)	.094
Type of amyloidosis				.707
Transthyretin	68 (88.3)	71 (91)	70 (89.7)	
Light chain	8 (10.4)	7 (9)	8 (10.3)	
Apo-A IV	1 (1.3)	0	0	
Hypertension	52 (67.5)	58 (74.4)	58 (74.4)	.345
Dyslipidemia	44 (57.1)	46 (59)	45 (57.7)	.946
Type 2 diabetes mellitus	17 (22.1)	18 (23.1)	17 (21.8)	.966
Former or current smoker	19 (24.7)	24 (30.8)	24 (30.8)	.627
Prior hospitalization due to heart failure	33 (42.9)	33 (42.3)	24 (30.8)	.122
Atrial fibrillation or flutter	50 (64.9)	48 (61.5)	26 (33.3)	< .001
Other arrhythmias	9 (11.7)	10 (12.8)	7 (9)	.591
Syncope	11 (14.3)	10 (12.8)	17 (21.8)	.205
Pacemaker implantation	13 (16.9)	13 (16.7)	9 (11.5)	.352
Ischemic heart disease	12 (15.6)	12 (15.4)	13 (16.7)	.854
Heart valve intervention	4 (5.2)	6 (7.7)	3 (3.8)	.712
Cerebrovascular disease	12 (15.6)	8 (10.3)	7 (9)	.200
Arterial or venous thrombosis	4 (5.2)	4 (5.1)	5 (6.4)	.742
Peripheral artery disease	5 (6.5)	6 (7.7)	2 (2.6)	.286
Chronic obstructive pulmonary disease	12 (15.6)	9 (11.5)	6 (7.7)	.126

Baseline characteristics

Variables	TAPSE/SPAP < 0.38 n = 77	TAPSE/SPAP 0.38 – 0.58 n = 78	TAPSE/SPAP > 0.58 n = 78	*P*
Clinical presentation				
Systolic blood pressure, mmHg	122.9 ± 17.1	126.7 ± 19.0	124.4 ± 16.0	.581
Diastolic blood pressure, mmHg	75 [65-82]	73.5 [64-80.5]	73 [63-82]	.283
Heart rate, bpm	72 [66-86]	76 [65-85]	69 [59-81]	.155
NYHA class III or IV	41 (53.2)	29 (37.2)	14 (17.9)	< .001
Exploratory signs of congestion	54 (70.1)	51 (65.4)	30 (38.5)	< .001
Laboratory				
Urea, mg/dL	75 [57-98]	67.5 [52.7-99.7]	69 [46.5-74.2]	.004
Creatinine, mg/dL	1.18 [0.97-1.56]	1.15 [0.99-1.48]	1 [0.83-1.25]	.725
Glomerular filtration rate, mL/min	51.6 [38.7-70.9]	51.3 [39.3-69.9]	70 [51.6-80.4]	.002
NT-proBNP, pg/mL[a]	4946 [2612-6863]	3292 [1682-6420]	1911 [627-3207]	< .001
UK NAC stages[a,b]*, %*				< .001
Stage I	17 (22.1)	30 (39.5)	48 (65.8)	
Stage II	37 (48.1)	24 (31.6)	20 (27.4)	
Stage III	23 (29.9)	22 (27.4)	5 (6.8)	
Potassium, mEq/L	4.3 [4.1-4.7]	4.4 [4.1-4.8]	4.5 [4.2-4.9]	.030
Sodium, mEq/L	140 [138-142]	140.5 [139-134]	141 [139-142]	.098
Hemoglobin, g/dL	13.6 [12.1-15.1]	13.4 [12.1-14]	13.5 [12.7-15.1]	.746
Uric acid, mg/dL	7.9 [6.4-9.4]	7.2 [5.8-8.3]	6.4 [5.2-7.5]	< .001
Bilirubin, mg/dL[a]	1 [0.8-1.3]	0.8 [0.6-1.1]	0.8 [0.6-1]	.129
Gamma-glutamyl transpherase, UI/L	90 [47.5-186]	54 [25.2-125.7]	37 [18-67.5]	.001
Alkaline phosphatase, UI/L	156 [110-208.5]	113.5 [83.7-190.2]	97 [68.7-149.7]	< .001
Albumin, g/dL[a]	4.1 [4.0-4.4]	4.1 [3.9-4.4]	4.1 [3.8-4.4]	.507
Total cholesterol, mg/dL	155.8 ± 41.3	153.1 ± 36.7	157.2 ± 38.0	.820

Baseline characteristics

Variables	TAPSE/SPAP < 0.38 n = 77	TAPSE/SPAP 0.38 – 0.58 n = 78	TAPSE/SPAP > 0.58 n = 78	*P*
Echocardiography				
LVEF, %	45.3 ± 10.2	53.8 ± 12.1	57.0 ± 11.9	< .001
TAPSE, mm	14 [12-15.8]	17 [15-19]	20 [17-22]	< .001
SPAP, mmHg	48.1 [41-54.6]	35.8 [30-40.7]	24.3 [21-28.2]	< .001
Maximum LV wall thickness, mm	17.2 [15.2-20]	17 [14-18]	17 [15-18]	.109
Moderate or severe aortic stenosis, %	8 (10.4)	10 (12.8)	8 (10.3)	.977
Moderate or severe mitral regurgitation, %	23 (30.7)	20 (26)	12 (15.4)	.027
Moderate or severe tricuspid regurgitation, %	38 (49.4)	22 (28.2)	5 (6.4)	< .001
Medications (at baseline) %				
Antiplatelet agents	8 (10.4)	14 (17.9)	17 (21.8)	.154
Anticoagulation	50 (64.9)	49 (62.8)	33 (42.3)	.004
Loop diuretics	69 (89.6)	70 (89.7)	45 (57.7)	< .001
Thiazides	10 (13)	6 (7.7)	10 (12.8)	.977
Beta-blockers	44 (57.1)	41 (52.6)	36 (46.2)	.172
ACEI or ARB or ARNI	35 (45.5)	35 (44.9)	39 (50)	.570
Mineralocorticoid antagonists	29 (37.7)	23 (29.5)	11 (14.1)	.001
Sodium-glucose cotransporter 2 inhibitors	11 (16.4)	7 (10.8)	6 (8.6)	.345
Other hypoglycemic agents	16 (20.8)	15 (19.2)	15 (19.5)	.840
Lipid-lowering agents	38 (49.4)	43 (55.1)	48 (61.5)	.128
Specific therapies (over follow-up) %				
Tafamidis	20 (26)	18 (23.1)	19 (24.4)	.816
Chemotherapy	7 (9.1)	5 (6.4)	7 (9)	.981
Autologous stem cell transplantation	2 (2.6)	0	4 (5.1)	.318

Clinical outcomes

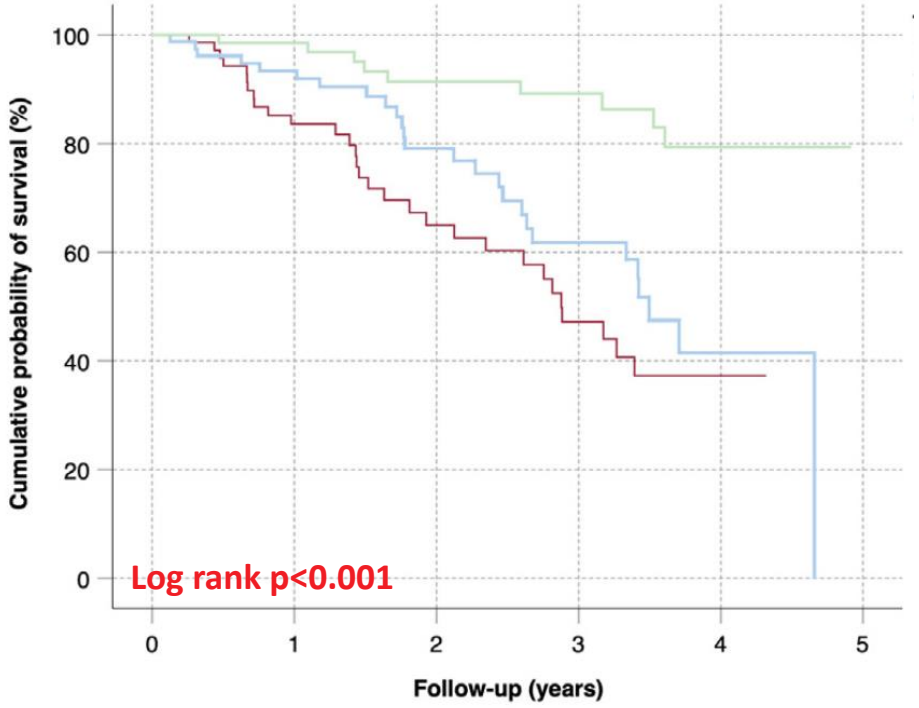
Median Follow-up = **680 days** (IQR 371–1234 days)

- **All-cause Death**=65 (27.9%)
- **CV Death**=47 (72.3%)
- **≥1 HF Hosp**=68 (29.2%)
- **HTx**=7 (3%)

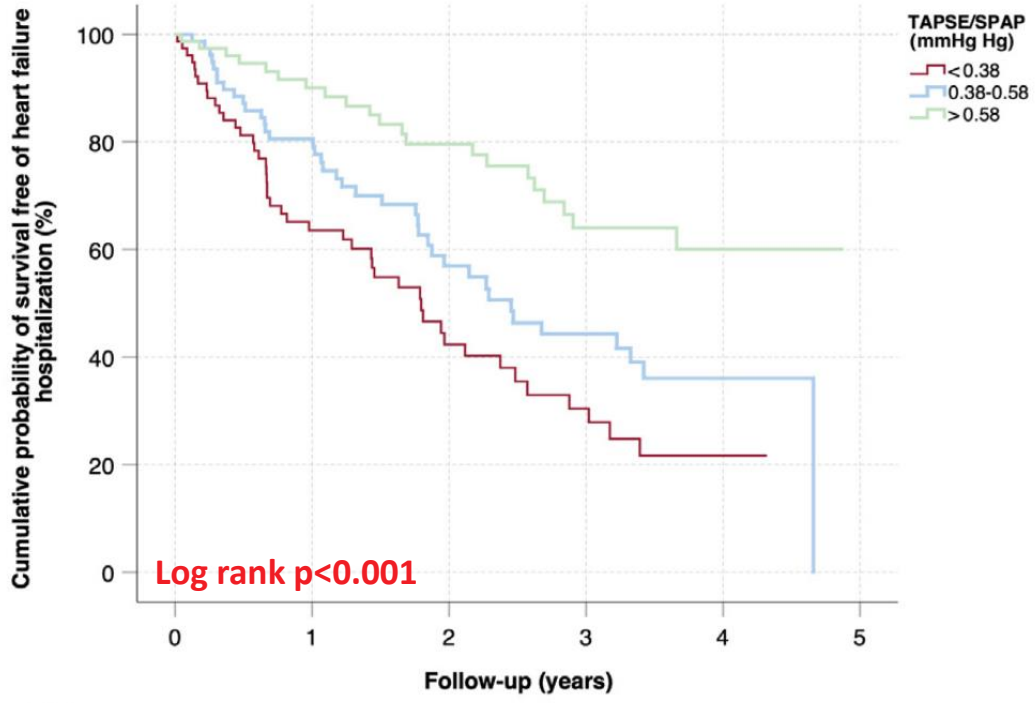
Causes of death	TAPSE/SPAP < 0.38 n = 30 deaths	TAPSE/SPAP 0.38 – 0.58 n = 26 deaths	TAPSE/SPAP > 0.58 n = 9 deaths
Cardiovascular, %	22 (73.3)	19 (73.1)	6 (66.7)
Heart failure	11	11	4
Sudden death	6	6	0
Noncerebral arterial embolism	2	0	1
Stroke	1	1	0
Unknown cause	2	1	1
Noncardiovascular, %	8 (26.7)	7 (26.9)	3 (33.3)
Infection	4	3	2
Malignancy	3	1	0
Other	1	3	1

SPAP, systolic pulmonary artery pressure; TAPSE, tricuspid annulus plane systolic excursion.
The data are presented as absolute numbers or No. (%).

Clinical Outcomes



Median survival (years)	Number at risk						
2.8	< 0.38	77	52	28	18	4	0
3.5	0.38-0.58	78	64	38	21	5	0
4.6	> 0.58	78	60	46	35	14	0



Number at risk						
<0.38	77	40	20	12	4	0
0.38-0.58	78	56	30	18	5	0
>0.58	78	57	41	26	11	0

Model	Death or heart failure hospitalization HR (95%CI)	Death HR (95%CI)
Univariable analysis		
TAPSE/SPAP ratio (per 0.1 mm/mmHg)	0.79 (0.72-0.87)	0.78 (0.68-0.89)
TAPSE/SPAP ratio < 0.38 vs > 0.58	3.36 (1.99-5.67)	5.44 (2.55-11.59)
TAPSE/SPAP ratio 0.38–0.58 vs > 0.58	2.22 (1.30-3.77)	3.72 (1.73-7.99)
Multivariable model 1 (clinical variables)		
TAPSE/SPAP ratio (per 0.1 mm/mmHg)	0.81 (0.73-0.89)	0.78 (0.68-0.89)
TAPSE/SPAP ratio < 0.38 vs > 0.58	3.11 (1.77-5.47)	6.25 (2.73-14.33)
TAPSE/SPAP ratio 0.38–0.58 vs > 0.58	1.98 (1.12-3.50)	4.26 (1.84-9.89)
Multivariable model 2 (laboratory variables)		
TAPSE/SPAP ratio (per 0.1 mm/mmHg)	0.82 (0.74-0.91)	0.80 (0.69-0.92)
TAPSE/SPAP ratio < 0.38 vs > 0.58	2.97 (1.69-5.23)	5.59 (2.41-12.96)
TAPSE/SPAP ratio 0.38–0.58 vs > 0.58	1.96 (1.09-3.51)	3.41 (1.44-8.04)
Multivariable model 3 (echo variables)		
TAPSE/SPAP ratio (per 0.1 mm/mmHg)	0.80 (0.72-0.90)	0.77 (0.66-0.89)
TAPSE/SPAP ratio < 0.38 vs > 0.58	3.00 (1.66-5.45)	6.27 (2.66-14.76)
TAPSE/SPAP ratio 0.38–0.58 vs > 0.58	2.19 (1.26-3.78)	4.22 (1.91-9.34)
Multivariable model 4 (drug variables)		
TAPSE/SPAP ratio (per 0.1 mm/mmHg)	0.79 (0.71-0.88)	0.76 (0.66-0.87)
TAPSE/SPAP ratio < 0.38 vs > 0.58	3.23 (1.85-5.64)	6.46 (2.90-14.38)
TAPSE/SPAP ratio 0.38–0.58 vs > 0.58	2.08 (1.18-3.66)	4.26 (1.89-9.62)
Multivariable model 5 (UK NAC staging system)		
TAPSE/SPAP ratio (per 0.1 mm/mmHg)	0.86 (0.78-0.96)	0.84 (0.73-0.97)
TAPSE/SPAP ratio < 0.38 vs > 0.58	2.11 (1.19-3.73)	4.06 (1.75-9.42)
TAPSE/SPAP ratio 0.38–0.58 vs > 0.58	1.57 (0.89-2.79)	3.08 (1.33-7.12)

	per 0.1 mm/mmHg	0.79 to 0.86	0.76 to 0.84
Multivariable HR	**<0.38 vs >0.58**	2.11 to 3.22	4.06 to 6.46
	0.38-0.58 vs >0.58	1.57 to 2.19	3.08 to 4.26

TAPSE/SPAP ratio and cardiac amyloidosis stages

UK NAC

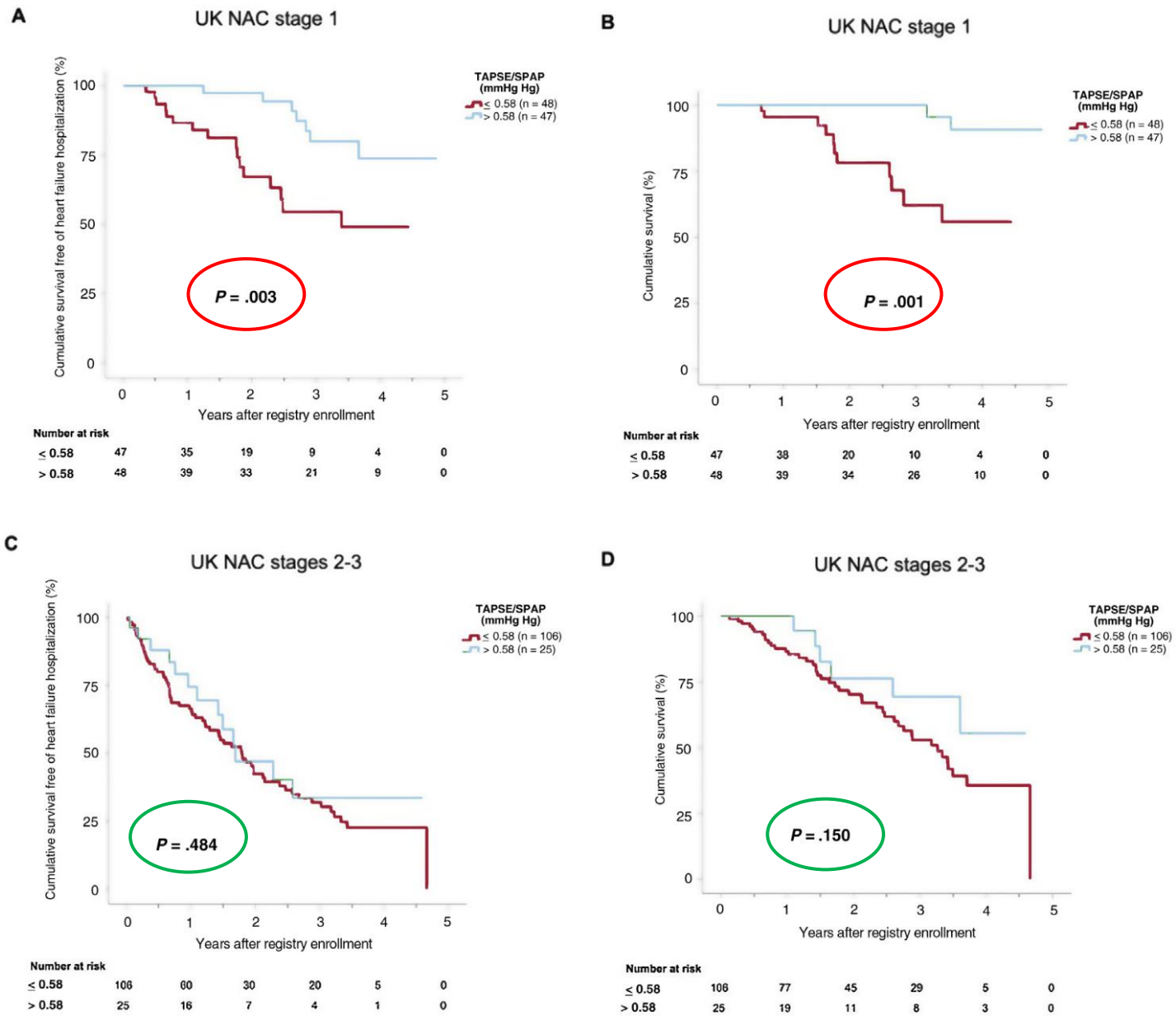
c-statistic all-cause death=0.662

c-statistic all-cause death or HFH=0.668

UK NAC + TAPSE/SPAP

c-statistic all-cause death=0.705 (p=0.065)

c-statistic all-cause death or HFH=0.707 (p=0.019)



Conclusions

- This study supports that the noninvasive and easy to obtain TAPSE/SPAP ratio is a strong and independent prognostic marker in patients with cardiac amyloidosis:

  Severe reduction -> **4-6 fold** increased mortality

  Mild-moderate reduction -> **3-4 fold** increased mortality

- This parameter may be additive to standard predictive systems based on biomarkers

- Our results highlight the important role of right ventricular to pulmonary afterload coupling in patients with cardiac amyloidosis

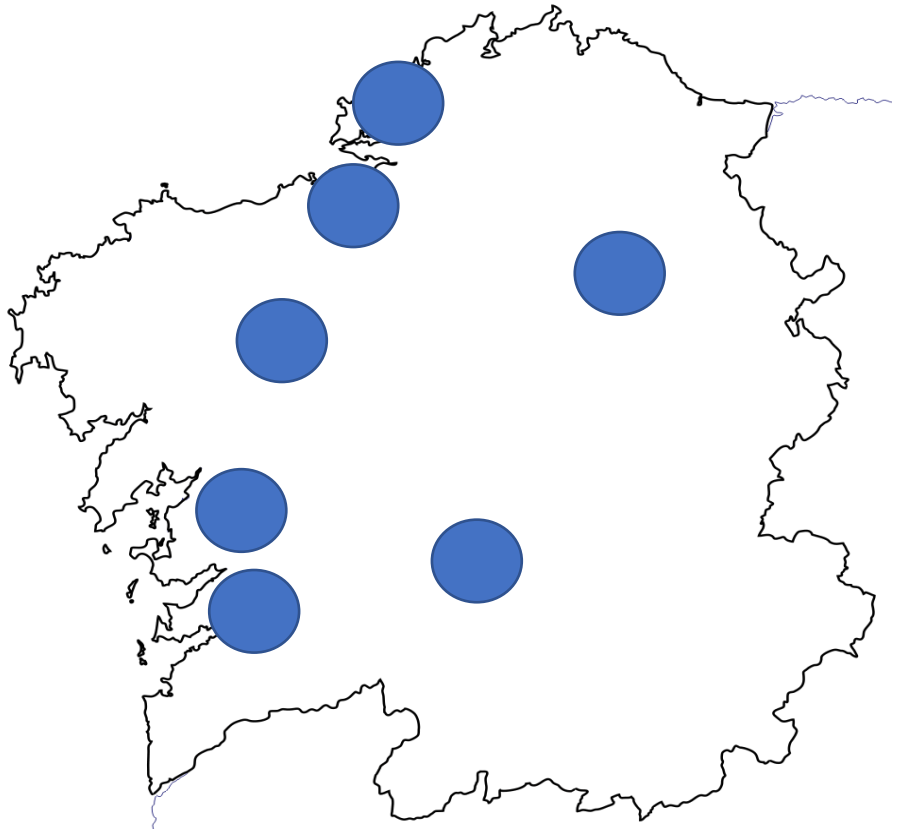
Registro de **AMI**loidosis cardiaca de **GAL**icia (AMIGAL)

PROSPECTIVE AND MULTICENTER REGISTRY

JANUARY 2018 – PRESENT

422 pacientes included

(31/12/2023)



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Original

Amiloidosis cardiaca: descripción de una serie de 143 casos

Gonzalo Barge-Caballero[a,b,*], Eduardo Barge-Caballero[a,b], Manuel López-Pérez[c], Raquel Bilbao-Quesada[d], Eva González-Babarro[e], Inés Gómez-Otero[b,f], Andrea López-López[g], Mario Gutiérrez-Feijoo[h], Alfonso Varela-Román[b,f], Carlos González-Juanatey[g], Óscar Díaz-Castro[d] y María G. Crespo-Leiro[a,b]

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INFORMACIÓN DEL ARTÍCULO

Historia del artículo:
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Palabras clave:
Amiloidosis cardiaca
Transtirretina
Cadenas ligeras
Registro
Multicéntrico
Prospectivo

RESUMEN

Introducción y objetivos: Recientemente se han producido importantes avances en el diagnóstico y tratamiento de la amiloidosis cardiaca (AC). Nos propusimos realizar una descripción actualizada de sus 2 tipos más frecuentes: la AC por transtirretina (AC-ATTR) y la AC por cadenas ligeras (AC-AL).
Métodos: Registro prospectivo de pacientes diagnosticados de AC en 7 hospitales de Galicia entre el 1 de enero de 2018 y el 30 de junio de 2020. Se recogieron variables relativas a características clínicas, pruebas complementarias, supervivencia y causas de muerte.
Resultados: Se incluyeron de forma consecutiva 143 pacientes con AC, 128 AC-ATTR (89,5%) y 15 AC-AL (10,5%). La edad media fue de 79,6 ± 7,7 años y un 23,8% fueron mujeres. La mayoría de los pacientes con AC-ATTR se diagnosticaron de forma no invasiva (87,5%). En la exploración física, un 35,7, un 35 y un 7% de los pacientes presentaban el signo de Popeye, contractura de Dupuytren y macroglosia, respectivamente. La supervivencia a los 12 y 24 meses fue del 92,1 y el 76,2% en el grupo AC-ATTR, y del 78,6 y el 61,1% en el grupo AC-AL (p = 0,152). La causa de muerte fue cardiovascular en el 80,8% de la cohorte.
Conclusiones: La AC-ATTR puede ser diagnosticada en la mayoría de los casos de manera no invasiva y es la forma de AC más frecuente en la práctica clínica habitual. Además, parece observarse un aumento en la supervivencia a corto plazo de la AC que en parte podría deberse a los avances relacionados con su diagnóstico y tratamiento.

Cardiac amyloidosis: Description of a series of 143 cases

Keywords:
Cardiac amyloidosis
Transthyretin
Light chain
Registry
Multicenter
Prospective

ABSTRACT

Introduction and objectives: Recently, there have been important advances in the diagnosis and treatment of cardiac amyloidosis (CA). Our aim was to provide an updated description of its 2 most frequent types: the transthyretin CA (ATTR-CA) and the light chain CA (AL-CA).
Methods: Prospective registry of patients with CA diagnosed in 7 institutions in Galicia (Spain) between January 1, 2018 and June 30, 2020. Variables related to clinical characteristics, complementary tests, survival and causes of death were collected.

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ORIGINAL ARTICLE

Beta-Blocker Exposure and Survival in Patients With Transthyretin Amyloid Cardiomyopathy

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Abstract

Objective: To investigate a potential association between beta-blocker exposure and survival in patients with transthyretin amyloid cardiomyopathy (ATTR-CM).
Methods: In this real-world prospective registry of 128 consecutive patients with ATTR-CM recruited in 7 institutions in Galicia (Spain), survival of 65 patients who received beta blockers on registry enrollment was compared with that of 63 untreated controls by means of both unweighted Cox regression and Cox regression with inverse probability of treatment weighting. Tolerance to and adverse effects of beta blockers were recorded. Median study follow-up was 520 days.
Results: Patients with ATTR-CM who received beta blockers showed statistically significant lower all-cause mortality than untreated controls as evaluated by either unweighted Cox regression (hazard ratio, 0.31; 95% CI, 0.12 to 0.79) or Cox regression with inverse probability of treatment weighting (hazard ratio, 0.18; 95% CI, 0.08 to 0.41; P<.001). Several sensitivity analyses confirmed the internal validity of these results. The overall frequency of beta-blocker suspension due to adverse effects was 25% (95% CI, 15.5% to 34.5%).
Conclusion: In this real-world, prospective, multi-institutional registry, patients with ATTR-CM who received beta blockers had lower all-cause mortality than untreated controls.

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Heart failure (HF) is the most frequent clinical manifestation of amyloid cardiomyopathy, and it is associated with poor outcomes.[1] In these patients, the prescription of neurohormonal blocking agents is a matter of concern as they may be poorly tolerated. Because of this, potential disease-modifying therapies are often denied to these individuals.

Current expert consensus documents advise against the routine prescription of beta blockers in patients with amyloid cardiomyopathy.[2,3] In the presence of advanced restrictive physiology, excessive bradycardia may lead to a reduced cardiac output, hypotension, fatigue, and dizziness. Neurogenic orthostatic intolerance, which is characteristic of some types of the disease, may also be aggravated.

However, a recent Italian single-center study[4] challenged this classic paradigm as it suggested that beta blockers might be initiated and up-titrated safely in a substantial proportion of patients with cardiac amyloidosis. Tolerance to beta blockers is better in patients with transthyretin amyloid cardiomyopathy (ATTR-CM) than in patients with light-chain cardiomyopathy,[4] a fact

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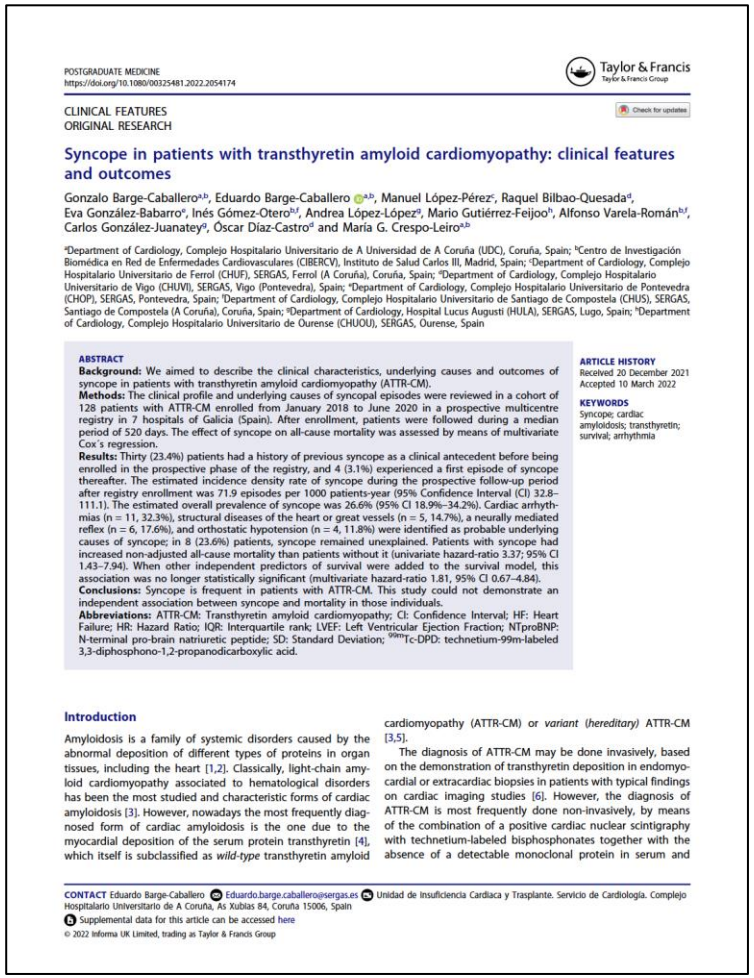

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CLINICAL FEATURES
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Syncope in patients with transthyretin amyloid cardiomyopathy: clinical features and outcomes

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ABSTRACT

Background: We aimed to describe the clinical characteristics, underlying causes and outcomes of syncope in patients with transthyretin amyloid cardiomyopathy (ATTR-CM).
Methods: The clinical profile and underlying causes of syncopal episodes were reviewed in a cohort of 128 patients with ATTR-CM enrolled from January 2018 to June 2020 in a prospective multicentre registry in 7 hospitals of Galicia (Spain). After enrollment, patients were followed during a median period of 520 days. The effect of syncope on all-cause mortality was assessed by means of multivariate Cox's regression.
Results: Thirty (23.4%) patients had a history of previous syncope as a clinical antecedent before being enrolled in the prospective phase of the registry, and 4 (3.1%) experienced a first episode of syncope thereafter. The estimated incidence density rate of syncope during the prospective follow-up period after registry enrollment was 71.9 episodes per 1000 patients-year (95% Confidence Interval (CI) 32.8–111.1). The estimated overall prevalence of syncope was 26.6% (95% CI 18.9%–34.2%). Cardiac arrhythmias (n = 11, 32.3%), structural diseases of the heart or great vessels (n = 5, 14.7%), a neurally mediated reflex (n = 6, 17.6%), and orthostatic hypotension (n = 4, 11.8%) were identified as probable underlying causes of syncope; in 8 (23.6%) patients, syncope remained unexplained. Patients with syncope had increased non-adjusted all-cause mortality than patients without it (univariate hazard-ratio 3.37; 95% CI 1.43–7.94). When other independent predictors of survival were added to the survival model, this association was no longer statistically significant (multivariate hazard-ratio 1.81, 95% CI 0.67–4.84).
Conclusions: Syncope is frequent in patients with ATTR-CM. This study could not demonstrate an independent association between syncope and mortality in those individuals.
Abbreviations: ATTR-CM: Transthyretin amyloid cardiomyopathy; CI: Confidence Interval; HF: Heart Failure; HR: Hazard Ratio; IQR: Interquartile rank; LVEF: Left Ventricular Ejection Fraction; NTproBNP: N-terminal pro-brain natriuretic peptide; SD: Standard Deviation; ^{99m}Tc-DPD: technetium-99m-labeled 3,3-diphosphono-1,2-propanodicarboxylic acid.

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KEYWORDS
Syncope; cardiac amyloidosis; transthyretin; survival; arrhythmia

Introduction

Amyloidosis is a family of systemic disorders caused by the abnormal deposition of different types of proteins in organ tissues, including the heart [1,2]. Classically, light-chain amyloid cardiomyopathy associated to hematological disorders has been the most studied and characteristic forms of cardiac amyloidosis [3]. However, nowadays the most frequently diagnosed form of cardiac amyloidosis is the one due to the myocardial deposition of the serum protein transthyretin [4], which itself is subclassified as *wild-type* transthyretin amyloid cardiomyopathy (ATTR-CM) or *variant* (*hereditary*) ATTR-CM [3,5].

The diagnosis of ATTR-CM may be done invasively, based on the demonstration of transthyretin deposition in endomyocardial or extracardiac biopsies in patients with typical findings on cardiac imaging studies [6]. However, the diagnosis of ATTR-CM is most frequently done non-invasively, by means of the combination of a positive cardiac nuclear scintigraphy with technetium-labeled bisphosphonates together with the absence of a detectable monoclonal protein in serum and

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Original

Incidencia y causas de hospitalización en pacientes con amiloidosis cardiaca por transtiretina (AC-ATTR) y por cadenas ligeras (AC-AL)

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Hospitalizaciones
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RESUMEN

Introducción y objetivos: La amiloidosis cardiaca (AC) es una patología asociada a un elevado número de ingresos hospitalarios. Dada la escasa información disponible al respecto, planteamos un análisis de la incidencia y las causas de hospitalización en esta enfermedad.
Material y métodos: Se evaluaron 143 pacientes (128 por transtiretina [AC-ATTR] y 15 por cadenas ligeras [AC-AL]) incluidos en el Registro de Amiloidosis Cardiaca de Galicia (AMIGAL), recogiendo todas sus hospitalizaciones.
Resultados: Durante un seguimiento mediano de 959 días se produjeron 179 hospitalizaciones no programadas (tasa de incidencia [TI] 512,6 ingresos hospitalarios por 1.000 pacientes-año), siendo las más habituales las de causa cardiovascular (n = 109, TI 312,2). El motivo individual de ingreso hospitalario más frecuente fue la insuficiencia cardiaca (IC) (n = 87, TI 249,2).
La AC-AL se asoció con una TI de hospitalizaciones no programadas más elevada que la AC-ATTR (TI 781 vs. 483,2; HR 1,62; p = 0,029) a expensas de las de causa no cardiovascular (TI 376 vs. 181,2; HR 2,07; p = 0,027). La supervivencia libre de hospitalización no programada al año y a los tres años en la AC-AL fue menor que en la AC-ATTR (46,7 y 20,0% vs. 73,4 y 35,2%, respectivamente; p = 0,021).
Conclusiones: La AC se asoció con una elevada incidencia de hospitalizaciones, siendo la causa individual más frecuente la IC; la supervivencia libre de hospitalización no programada en la AC-AL fue menor que en la AC-ATTR, debido principalmente a los ingresos hospitalarios de causa no cardiovascular.

Keywords:
Cardiac amyloidosis
Transthyretin amyloidosis

Incidence and causes of hospitalization in patients with transthyretin (ATTR-CA) and light chain (AL-CA) cardiac amyloidosis

ABSTRACT

Introduction and objetives: Cardiac amyloidosis (CA) is a disorder associated with high number of hospital admissions. Given the scarce information available, we propose an analysis of the incidence and causes of hospitalization in this disease.

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Original article

Prognostic value of the tricuspid annular plane systolic excursion/systolic pulmonary artery pressure ratio in cardiac amyloidosis

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ABSTRACT

Introduction and objectives: The tricuspid annular plane systolic excursion/systolic pulmonary artery pressure (TAPSE/SPAP) ratio is a noninvasive surrogate of right ventricular to pulmonary circulation that has prognostic implications in patients with heart failure (HF) or pulmonary hypertension. Our purpose was to evaluate the prognostic value of the TAPSE/SPAP ratio in patients with cardiac amyloidosis.
Methods: We used the database of the AMIGAL study, a prospective, observational registry of patients with cardiac amyloidosis recruited in 7 hospitals of the Autonomous Community of Galicia, Spain, from January 1, 2018 to October 31, 2022. We selected patients whose baseline TAPSE/SPAP ratio was calculated with transthoracic echocardiography. Long-term survival and survival free of HF hospitalization were assessed by means of 5 different multivariable Cox regression models. Median follow-up was 680 days.
Results: We studied 233 patients with cardiac amyloidosis, among whom 209 (89.7%) had transthyretin type. The baseline TAPSE/SPAP ratio correlated significantly with clinical outcomes. Depending on the multivariable model considered, the adjusted hazard ratios estimated per 0.1 mm/mmHg increase of baseline TAPSE/SPAP ratio ranged from 0.76 to 0.84 for all-cause mortality. Similarly, the ratios for all-cause mortality of HF hospitalization ranged from 0.79 to 0.84. The addition of the baseline TAPSE/SPAP ratio to the predictive model of the United Kingdom National Amyloidosis Centre resulted in an increase in Harrell's c-statistic from 0.662 to 0.705 for all-cause mortality and from 0.668 to 0.707 for all-cause mortality or HF hospitalization.
Conclusions: Reduced TAPSE/SPAP ratio is an independent adverse prognostic marker in patients with cardiac amyloidosis.

Valor pronóstico de la razón desplazamiento sistólico del plano del anillo tricuspídeo/presión arterial pulmonar sistólica en la amiloidosis cardiaca

Palabras clave:
Amiloidosis cardiaca
Desplazamiento sistólico del plano del anillo tricuspídeo
Presión arterial pulmonar sistólica
Función del ventrículo derecho
Acoplamiento ventriculoarterial

RESUMEN

Introducción y objetivos: La razón entre el desplazamiento sistólico del plano del anillo tricuspídeo y la presión arterial pulmonar sistólica (TAPSE/PAPS) es una medida no invasiva del acoplamiento entre el ventrículo derecho y la circulación pulmonar con implicaciones pronósticas en pacientes con insuficiencia cardiaca (IC) o hipertensión pulmonar. El objetivo es evaluar el valor pronóstico del cociente TAPSE/PAPS en pacientes con amiloidosis cardiaca.

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Thank you

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