

## SUPPLEMENTARY DATA

**Table S1.** Bone scintigraphy imaging

Grading	Value
Perugini grade 0	44 (10.0)
Perugini grade 1	40 (9.0)
Perugini grade 2	26 (5.9)
Perugini grade 3	210 (47.5)
Not performed	89 (20.1)
Missing	33 (7.5)
Data are expressed as no. (%).	

**Table S2.** Type of mutation (N = 442)

Mutation	Value
p.Val30Met	287 (64.9)
p.Val122Ile	95 (21.5)
p.Glu89Lys	23 (5.2)
p.Ser97Tyr	13 (2.9)
p.Ala65Thr	5 (1.1)
p.Val122Del	4 (0.9)
p.Ser43Asn	3 (0.7)
p.Leu32Val	3 (0.7)
p.Asp38Asn	2 (0.5)
p.Glu89Gln	2 (0.5)
p.Val71Ala	1 (0.2)
p.Glu74Gln	1 (0.2)
p.Ala128Val	1 (0.2)
p.Ser50Arg	1 (0.2)
p.Thr69Ile	1 (0.2)

Data are expressed as No. (%).



**Table S3.** Comparison of groups according to treatment status (treated and untreated with tafamidis, patisiran, and/or inotersen)

Characteristic	Untreated		Treated <sup>a</sup>		<i>P</i>
	Value	No. <sup>b</sup>	Value	No. <sup>b</sup>	
<i>Age</i> ≥ 80 y	32 (22.4)	143	6 (3.2)	187	< .001 <sup>a</sup>
<i>Male sex</i>	77 (53.1)	145	149 (75.3)	198	< .001 <sup>a</sup>
<i>FEV<sub>1</sub></i> ≤ 50 %	36 (27.1)	133	39 (21.2)	184	.280 <sup>a</sup>
<i>NYHA</i>					
I+II	119 (86.9)	137	182 (92.9)	196	.101 <sup>a</sup>
III+IV	278 (86.3)	137	298 (92.5)	196	

FEV<sub>1</sub>, forced expiratory volume in 1 second; NYHA, New York Heart Association functional class.

<sup>a</sup>Chi-square test.

<sup>b</sup>No. number of patients with available data. Data are expressed as No. (%).

**Table S4.** Death case numbers by group

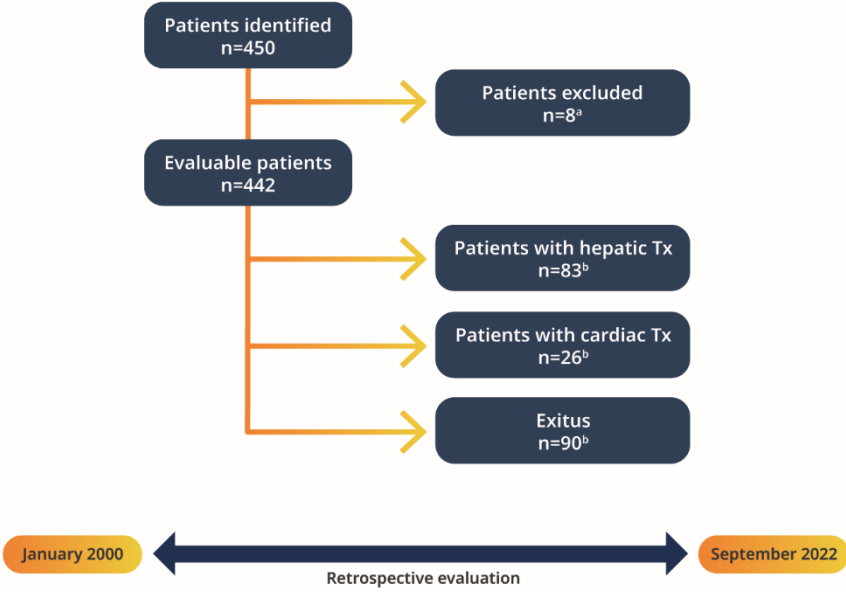
Characteristic	Value		Value	<i>P</i> <sup>a</sup>
p.Val30Met [261]	55 (21.1)	Other mutation [n = 150]	35 (23.3)	.682
p.Val122Ile [91]	24 (26.4)	Other mutation [n = 320]	66 (20.6)	.305
p.Glu89Lys [23]	4 (17.4)	Other mutation [n = 388]	86 (22.2)	.781
Neuropathy [262]	63 (24.0)	Non-neuropathy [n = 148]	26 (17.6)	.160
Treated <sup>b</sup> [259]	41 (15.8)	Untreated [n = 141]	44 (31.2)	< .001

Data are expressed as No. (%). The number of patients with available data is expressed in brackets.

<sup>a</sup>Chi-square test.

<sup>b</sup>Treated and untreated with tafamidis, patisiran, inotersen and/or liver transplantation

Figure S1. Patient characteristics



Correcciones a la figura S1.

Poner espacio a ambos lados de “=”

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Figure S2. Number of patients provided per center

SITE	REGION	CITY	N
H.U. Son Llàtzer	Balearic Islands	Palma de Mallorca	54
H.U. Puerta de Hierro	Community of Madrid	Madrid	49
H.U. Bellvitge	Catalonia	Barcelona	42
H.U. Juan Ramón Jiménez	Andalusia	Huelva	36
H.U. La Fe	Valencian Community	Valencia	32
H.U. Son Espases	Balearic Islands	Palma de Mallorca	23
H.U. Virgen de la Victoria	Andalusia	Málaga	18
H.U. Araba - Txagorritxu	Basque Country	Vitoria	17
H.U. Vall d'Hebron	Catalonia	Barcelona	14
Hospital Clínic	Catalonia	Barcelona	12
C.A.U. de Salamanca	Castile and Leon	Salamanca	11
H.G.U. Gregorio Marañón	Community of Madrid	Madrid	10
H.U. Reina Sofía	Andalusia	Córdoba	10
H.U. Virgen de las Nieves	Andalusia	Granada	10
H.U. 12 de Octubre	Community of Madrid	Madrid	9
H.U. Donostia	Basque Country	Guipúzcoa	9
H.U. Virgen de la Arrixaca	Region of Murcia	Murcia	8
Hospital Marina Salud	Valencian Community	Valencia	8
H.U. de Jaén	Andalusia	Jaén	7
C.H.U. de A Coruña	Galicia	A Coruña	7
H.U. Josep Trueta	Catalonia	Girona	6
H.U. Dr Negrín	Canary Islands	Gran Canaria	7
H.U. Arnau de Vilanova	Catalonia	Lleida	6
Hospital Clínico San Carlos	Community of Madrid	Madrid	5
H.U. Basurto (Bilbao)	Basque Country	Vizcaya	5
H.U. Marqués de Valdecilla	Cantabria	Cantabria	5
H.C.U. Lozano Blesa	Aragon	Zaragoza	4
H.H.U. de Galdakao	Basque Country	Vizcaya	4
H.U. Puerto Real	Andalusia	Cádiz	4
H.U. Virgen del Rocío	Andalusia	Sevilla	4
H.U. Ramón y Cajal	Community of Madrid	Madrid	4
H.G.U. de Elche	Valencian Community	Alicante	3
H.U. Ntra. Sra. de Candelaria	Canary Islands	Tenerife	3
H. U. de Navarra	Navarra	Pamplona	3
H.G.U. de Alicante	Valencian Community	Alicante	2
H.C.U. de Valladolid	Castile and Leon	Valladolid	2
H. del Mar	Catalonia	Barcelona	1
H. Álvaro Cunqueiro	Galicia	Pontevedra	1
H. U. Toledo	Castile-La Mancha	Toledo	1
<b>Total</b>			<b>456</b>

C.A.U., Complejo Asistencial Universitario; C.H.A., Complejo Hospitalario Universitario; H.C.U., Hospital Clínico Universitario; H.G.U., Hospital General Universitario; H.U., hospital Universitario



**Correcciones a la figura S2:**

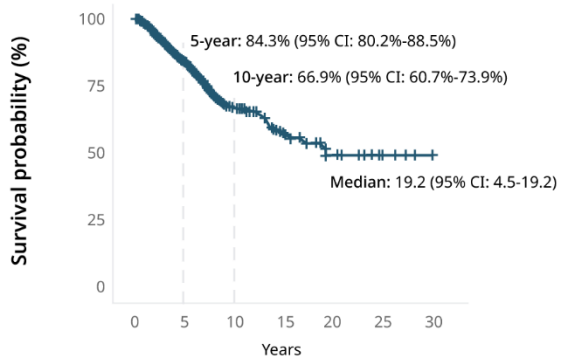
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**Cambiar “Navarra” a “Navarre”**

**Cambiar “N” a “No.” En la tabla**

**Figure S3. Survival**

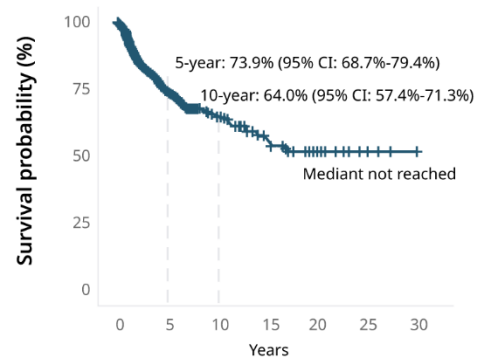
A)



Number at risk

All patients 374 198 79 42 20 9 2

B)



Number at risk

All patients 404 127 58 28 17 8 4

Correcciones a la figura S3

Cerrar "95%CI"

Cambiar "Mediant" a "Median"