

## **Role of natriuretic peptide to predict cardiac abnormalities in patients with hereditary transthyretin amyloidosis.**

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**Table 1: Baseline clinical, biological, and treatment characteristics of subjects according to phenotype.**

Phenotypes	Asymptomatic	Neurological	Cardiac with or without neurological	p
<b>N</b>	4	6	26	
<b>Genetic mutation</b>				
<i>ATTR</i> Val30Met, <i>n</i> (%)	3 (80)	5 (83)	11 (42)	na
<b>Clinical variables</b>				
Age, years	48 (33; 56)	40 (35; 49)	65 (56; 74)	0.006
Weight, kg	73 (63; 80)	59 (53; 72)	71 (57; 75)	0.32
BMI, kg/m <sup>2</sup>	24 (22; 25)	24 (22; 28)	23 (20; 26)	0.58
Men, <i>n</i> (%)	3(75)	0	20(77)	0.002
SBP, mmHg	140 (136; 162)	110 (105; 132)	120 (110; 130)	0.045
DBP, mmHg	80 (75; 97)	70 (63; 86)	80 (70; 80)	0.59
HR, bpm	74 (69; 91)	80 (69; 86)	80 (69; 86)	0.39
NYHA class (3; 4), %	0	0	2 (8)	0.67
History of decompensated HF	0	0	5 (19)	0.27
Carpal-tunnel syndrome or history of decompression, %	0	0	14 (54)	0.018
Peripheral neuropathy, <i>n</i> (%)	0	6(100)	21(81)	0.001
<b>Dysautonomia</b>				
Gastric and gut, <i>n</i> (%)	0	4 (67)	15 (58)	0.08
Orthostatic hypotension, <i>n</i> (%)	0	0	5 (19)	0.33
Erectile dysfunction, <i>n</i> (%)	0	0	13 (50)	0.02
Urinary dysfunction, <i>n</i> (%)	0	1 (17)	2 (8)	0.63
<b>Electrocardiogram</b>				
PR interval, (ms)	144 (123; 175)	179 (162; 198)	190 (146; 220)	0.19
QRS interval, (ms)	85 (80; 101)	96 (76; 109)	98 (80; 120)	0.42
<b>Treatment</b>				
Liver transplant, <i>n</i> (%)	0	3 (50)	5 (19)	0.33
Cardiac-pacemaker implant, <i>n</i> (%)	0	0	14 (56)	0.007
<b>Biological variables</b>				
NT- proBNP, pg/ml	33 (19; 50)	54 (37; 154)	747 (253; 2840)	0.001
cTnT, ng/ml	0.01	0.01	0.24 (0.01; 0.05)	0.02
Creatinine, μmol/l	82 (67; 92)	71 (61; 100)	83 (69; 101)	0.69
Hemoglobin, g/dl	14.4 (13.2; 15.7)	12.7 (12.1; 13.6)	12.7 (12.0; 14.1)	0.21

BMI: body-mass index; SBP: systolic blood pressure; DBP: diastolic blood pressure; HR: heart rate.

**Table 2: Baseline echocardiographic characteristics in subjects according to phenotype.**

<b>Phenotype:</b>	<b>Asymptomatic</b>	<b>Neurological</b>	<b>Cardiac with or without neurological</b>	
<b>Echocardiographic variables</b>				
<b>N</b>	4	6	26	
<b>LV variables</b>				
LVEDD, mm	50 (48; 53)	47 (41; 49)	43 (40; 49)	0.14
LVESD, mm	30 (28; 31)	29 (23; 30)	29 (25; 35)	0.61
IVST, mm	9 (8; 9)	9 (8; 10)	16 (12; 19)	0.0001
LVPWTD, mm	8 (7; 9)	9 (8; 9)	14 (10;16)	0.0001
LVEF, mm	61 (59; 73)	65 (60; 67)	60 (46; 63)	0.20
LV 2D strain, %	-19 (-17; -20)	-19 (-18; -22)	-12 (-10; -15)	0.004
LVM, g	143 (126; 178)	141 (125; 152)	258 (213; 383)	0.002
LVMind, g/m <sup>2</sup>	79 (68; 90)	93 (70; 95)	143 (119; 217)	0.001
E, cm	163 (129; 180)	75 (73; 84)	74 (59; 104)	0.64
A, cm	61 (46; 75)	57 (45; 77)	58 (39; 74)	0.63
E/A	1.5 (1.0; 1.7)	1.3 (1.1; 1.7)	1.3 (0.8; 2.5)	0.98
E/Ea	6 (6; 7)	7 (7; 9)	12 (6; 20)	0.14
<b>Other variables</b>				
Ao, mm	30 (26; 30)	28 (24; 31)	32 (29;36)	0.025
LA, mm	37 (30; 38)	34 (28; 38)	40 (35; 47)	0.042
SPAP, mmHg*	20	20	41(27; 59)	0.24
TAPSE, mm	26 (25; 27)	19 (18; 23)	17 (13; 22)	0.023
Pericardial effusion, %	0	1 (17)	9 (35)	0.29

LVEDD: left ventricular end-diastolic diameter; IVST: inter-ventricular septal thickness, and LVPWT: left ventricular posterior wall thickness; LV 2D strain: LV systolic function; LVM; LVM: left ventricular mass as marker of amyloid deposits; LVMind: LVM indexed by the body surface area; E: peak of early transmitral wave in pulsed Doppler; A: peak of atrial transmitral wave in pulsed wave Doppler; Ea: mean of early septal and lateral diastolic peak measured in tissue Doppler imaging; Ao: aortic diameter; LA: left atria diameter; SPAP: systolic pulmonary artery pressure; TAPSE: tricuspid annular plane systolic excursion. \*Only measureable in patients with tricuspid regurgitation ( $n=16$ ).