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ORIGINAL ARTICLE

Right ventricular involvement in transthyretin amyloidosis

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ABSTRACT

Background: The extent of right ventricular (RV) involvement in transthyretin amyloidosis (ATTR) is unknown.

Objectives: This study sought to establish the degree of RV involvement in ATTR amyloidosis, and compare findings with RV involvement in hypertrophic cardiomyopathy (HCM).

Methods: Forty-two patients with ATTR amyloidosis and echocardiographic evidence of cardiac amyloidosis (cardiac ATTR), 19 ATTR patients with normal left ventricular (LV) wall thickness (non-cardiac ATTR), 25 patients with diagnosed HCM and 30 healthy controls were included in this study. Echocardiographic measurements for conventional parameters, as well as RV global and segmental strain, were recorded.

Results: When comparing RV structure and function between cardiac ATTR amyloidosis and HCM patients, only segmental strain differed between the two groups. In cardiac ATTR amyloidosis, we found an RV apex-to-base strain gradient with highest deformation in the apex. This pattern was reversed in patients with HCM.

Conclusions: RV involvement is common in cardiac ATTR patients. The present study also detected an RV apical sparing pattern in patients with ATTR cardiomyopathy, similar to what has previously been described for the left ventricle in these patients. This pattern was not seen in HCM patients. Further studies are needed to assess the clinical importance of these findings.

Abbreviations: A: late diastolic left ventricular filling; AL: amyloid light chain; ATTR: transthyretin amyloid; E: early diastolic left ventricular filling; e': peak early diastolic tissue velocity; FW: free wall; HCM: hypertrophic cardiomyopathy; IVRT: isovolumic relaxation time; IVST: interventricular septal thickness; LV: left ventricle; LVEDV: left ventricular end diastolic volume; PWT: posterior wall thickness; RA: right atrial; RV: right ventricle; RVEDA: RV end diastolic area; RVEDD: end-diastolic RV diameter; RVESA: RV end systolic area; RVT: right ventricular thickness; RV s': right ventricular systolic tissue velocity; RV e': right ventricular early diastolic tissue velocity; RV a': right ventricular late diastolic tissue velocity; TAPSE: systolic displacement of the tricuspid annulus; TTR: transthyretin

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Introduction

Knowledge of right ventricular (RV) function has become increasingly important as RV function has recently been shown to predict survival and exercise performance in heart failure (HF) [1]. RV structure is significantly different from that of the left ventricle (LV) [2]. The RV is composed of two compartments, the inflow and the outflow tracts [3,4], which are different, both structurally and in the way they are interrelated, compared to the same structures of the LV. The myocardial fiber arrangement of the two ventricles also differs significantly, explaining partly the difference in functional patterns between the LV and RV. In addition, cavity wall thickness differentiates the two ventricles; the RV walls being thinner compared to the walls of the LV, in accordance with the pressures in the circulations the chambers support [5].

It has been shown that right heart function is affected in different types of chronic heart failure including idiopathic and ischemic cardiomyopathies [6,7] as well as in different causes of increased LV thickness [8,9]. In patients with increased LV afterload, RV structure and function may look normal; however, in those with more generalized cardiac myopathy, for example, hypertrophic cardiomyopathy (HCM), RV walls may exhibit hypertrophic thickening along with reduced strain [10]. In immunoglobulin light chain (AL) amyloidosis RV involvement is relatively common [11], and when present depicts a poor prognosis [12,13]. Conversely, RV involvement in hereditary transthyretin (ATTR) amyloidosis has been sparsely investigated, and to the best of our knowledge, no previous study has investigated regional RV function in detail.

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Therefore, we aimed to investigate the degree of RV involvement in ATTR amyloidosis patients, and compare our findings with RV parameters and findings from HCM patients, as both diseases are examples of pathologies causing ventricular wall thickening.

Patients and methods

Sixty-five patients, with tissue biopsy and genetically diagnosed ATTR amyloidosis, were randomly selected from ATTR patients undergoing clinical evaluation at Umeå University Hospital between 2005 and 2014 [14]. All patients had undergone a transthoracic echocardiographic examination and were reported as having cardiac amyloid involvement “cardiac ATTR” if the interventricular septal thickness (IVST) was >12 mm. Patients with IVST ≤ 12 mm were categorized as ‘non-cardiac ATTR’ [15]. Four ATTR amyloidosis patients were excluded from the analysis; 3 due to atrial fibrillation and 1 due to a significant atrial septal defect resulting in RV volume overload. Therefore, a total of 61 ATTR patients, median age 64 years (range 57–72 years), were included in the study. Most patients carried the transthyretin (TTR) V30M ($n = 58$) genetic mutation; other genotypes were TTR G54L ($n = 1$), T60A ($n = 1$) and A45S ($n = 1$) mutations. Twenty-five patients with biopsy proven ($n = 16$) or genetically verified ($n = 9$) HCM diagnosis, median age 54 years (range 41–66 years) were also included in the study. These patients fulfilled the echocardiographic criteria for HCM (IVST ≥ 15 mm), and the extent of wall thickening seen could not be explained by increased loading conditions such as long-standing hypertension or aortic stenosis [16,17].

In addition, 30 healthy controls, median age 61 years (range 51–69 years), were studied. The control group comprised of a subset of individuals originally recruited to the Umeå General Population Heart Study [18]. None of the controls had any cardiovascular or systemic disease, and did not use any medications known to influence cardiac function. All parts of the study complied with the Declaration of Helsinki, written informed consent was obtained from study participants, and the study approved by the Regional Ethical Committee (Umeå, Sweden).

Echocardiographic analysis

Patients and controls underwent a comprehensive echocardiographic examination, including 2D, M-Mode, pulsed and tissue Doppler echocardiography using GE Vivid 7, E9 (GE Vingmed Ultrasound, Horten, Norway) or Philips IE 33 (Philips ultrasound, Bothell, WA). For both HCM and cardiac ATTR patients, the echocardiographic analysis was made off-line using commercially available software packages Echopac PC version 113, (GE Healthcare, Horten, Norway) and TomTec Imaging Systems, Version 4.5, (Unterschleissheim, Germany). All analysis was performed by one operator, SA.

Data analysis was conducted according to the recommendations of American Society of Echocardiography [19,20]; from the parasternal long axis view, end-diastolic IVST and posterior wall thickness (PWT) were measured. From the apical four-chamber view measurements of early (E) and late (A) diastolic LV filling velocities were acquired using pulsed

Doppler recordings, with the sample volume placed at the tips of the mitral valve leaflets, and E/A ratio was calculated. For further assessment of LV diastolic function, peak early diastolic tissue velocity (e') was measured from myocardial pulsed Doppler recordings with the sample volume placed at the basal segment of the lateral LV wall, E/e' was calculated and taken as an indirect reflection of left atrial pressures [21].

The right heart structure and function was also assessed from the apical 4-chamber view as previously described [22]. Right heart geometrical measurements including end-diastolic RV diameter (RVEDD), RV end diastolic (RVEDA) and end-systolic areas (RVESA) were also measured and RV fractional area change (RVFAC), as well as right atrial area (RA area) in end-systole calculated. When image quality was adequate, RV end-diastolic free wall thickness (RVT) was also measured from the subcostal view; a value of >5 mm was considered as increased RVT [20]. RV systolic function was assessed by M-mode measured RV systolic displacement of the tricuspid annulus (TAPSE) from RV free wall. TAPSE was defined as the total displacement between the time of the Q-wave of the ECG and end systole (end of T-wave on ECG); TAPSE <17 mm was a marker of impaired systolic RV longitudinal function [20]. From pulsed myocardial Doppler velocity recordings, with the sample volume positioned at the base of the RV free wall, peak systolic (RV s'), early diastolic (RV e') and late diastolic (RV a') velocities were measured as well as the isovolumic relaxation time (IVRT_{rv}). RV and right atrial (RA) pressures gradient were also estimated from peak systolic tricuspid regurgitation (RV–RA peak systolic gradient).

Myocardial deformation analysis

Echocardiographic examinations, saved in digital imaging and communications in medicine (DICOM) format, were exported to vendor independent TomTec imaging arena where speckle tracking strain analysis was carried out. From the apical four-chamber view, RV and LV longitudinal endocardial strain was determined from manually tracing the endocardial border of the end-systolic frame, including the IVST. The software algorithm thereafter automatically defined the endocardial border in subsequent frames throughout the cardiac cycle and divided the RV and LV into six segments. Automatic tracking quality was assessed and, if needed, was manually adjusted to ensure optimal tracking of each segment. Only segments with optimal speckle tracking for analysis were included. The resulting strain values were averaged for the six segments to generate a global peak RV and LV longitudinal strain. RV strain analysis was only executed when RV free wall was visible throughout the entire cardiac cycle. Calculation of both RV segmental, RV free wall (RV FW), and global peak systolic strain (RV global) were performed. In addition, potential apex-to-base gradient patterns, for both the RV and LV, were evaluated by applying the formula proposed by Phelan et al. at the RV [23].

Statistics

Statistical analysis was performed using commercially available software (IBM SPSS statistics, version 22). Dichotomous

Table 1. Demographic and clinical characteristics of patient groups and controls.

	Cardiac ATTR, <i>n</i> = 42	HCM, <i>n</i> = 25	<i>p</i> Value*	Non-cardiac ATTR, <i>n</i> = 19	Controls, <i>n</i> = 30	<i>p</i> Value†
Gender, M/F	28/14	12/13	.198	10/9	16/14	.596
Age, years	69 (63–74)	54 (41–66)	<.0001	49 (40–60)	61 (51–69)	.005
Height, cm	173 (167–180)	169 (162–187)	.833	173 (164–183)	173 (164–178)	.944
Weight, kg	70 (63–77)	85 (66–100)	.028	71 (64–81)	74 (67–82)	.494
Systolic BP, mmHg	135 (120–150)	133 (113–168)	.857	125 (118–133)	140 (111–149)	.113
Diastolic BP, mmHg	75 (70–85)	80 (76–96)	.125	75 (70–80)	78 (70–85)	.851

ATTR: transthyretin amyloidosis; BP: blood pressure; HCM: hypertrophic cardiomyopathy; M/F: males/females.

*Statistical differences between cardiac ATTR and HCM.

†Statistical differences between non-cardiac ATTR and controls.

Significant differences ($p < .05$) are marked in bold.**Table 2.** Echocardiographic findings in patient groups and controls.

	Cardiac ATTR (<i>n</i> = 42)	HCM (<i>n</i> = 25)	<i>p</i> Value*	Non-cardiac ATTR (<i>n</i> = 19)	Controls (<i>n</i> = 30)	<i>p</i> Value†
Heart rate, bpm	72 (62–80)	67 (60–75)	.127	82 (70–90)	63 (59–69)	<.0001
IVST, mm	16 (14–18)	18 (15–20)	.017	11 (9–11)	10 (9–11)	.075
PWT, mm	12 (10–13)	10 (9–12)	.004	8 (8–9)	9 (8–9)	.664
IVST/PWT	1.4 (1.2–1.5)	1.8 (1.6–2.1)	<.0001	1.2 (1.1–1.4)	1.1 (1.0–1.3)	.168
E/A	0.9 (0.7–1.2)	1.3 (0.9–1.5)	.028	1.0 (0.8–1.3)	1.0 (0.8–1.4)	.772
E/e'	8.9 (7.3–13.1)	9.9 (6.1–12.4)	.928	7.4 (6.0–9.4)	6.4 (5.3–7.3)	.119
RVEDD, mm	36 (31–41)	34 (31–40)	.732	34 (31–38)	37 (34–39)	.068
RA area, cm ²	15.6 (14.1–17.2)	15.0 (14.0–18.0)	.936	14.4 (12.5–15.8)	13.8 (12.8–16.9)	.982
Abnormal RVT >5 mm, <i>n</i> (%)	20 (57)	6 (35)	.237	1 (7)	0 (0)	–
TAPSE <17 mm, <i>n</i> (%)	10 (24)	2 (8)	.154	0 (0)	0 (0)	–
IVRT, ms	57 (39–89)	–	–	17 (0–59)	22 (3–33)	.958
RV s', cm/s	11.3 (9.7–14.0)	–	–	12.0 (10.8–14.9)	12.0 (11.3–13.7)	.485
RV e', cm/s	9.5 (6.9–13.2)	–	–	12.0 (8.0–14.0)	11.7 (10.0–13.4)	.718
RV a', cm/s	14.5 (10.6–17.2)	–	–	13.5 (10.6–17.4)	14.3 (12.6–15.7)	.644
RVEDA, cm ²	14.7 (12.3–18.0)	15.3 (14.8–20.9)	.300	12.9 (11.2–15.6)	15.8 (13.2–18.3)	.034
RVFAC, %	42 (38–48)	40 (39–46)	.260	43 (37–50)	41 (37–54)	.930
RV–RA peak systolic gradient, mm Hg	27 (17–31)	27 (21–29)	.920	23 (14–34)	24 (19–30)	.820

ATTR: transthyretin amyloidosis; E/A: early to late mitral diastolic velocities ratio; E/e': early diastolic mitral to early diastolic tissue velocities ratio; HCM: hypertrophic cardiomyopathy; IVRT: isovolumic relaxation time; IVST: interventricular septal thickness; LV: left ventricular; PWT: posterior wall thickness; RA: right atrial; RV: right ventricular; RVEDA: right ventricular end diastolic area; RVEDD: right ventricular end diastolic diameter; RVFAC: right ventricular fractional area change; RVT: right ventricular thickness; RV a': right ventricular late diastolic tissue velocity; RV e': right ventricular early diastolic tissue velocity; RV s': right ventricular systolic tissue velocity; TAPSE: systolic displacement of the tricuspid annulus.

*Statistical differences between cardiac ATTR and HCM.

†Statistical differences between non-cardiac ATTR and controls.

Significant differences ($p < .05$) are marked in bold.

data were presented as counts or percentages, and count data as medians and interquartile range (25–75 percentiles), unless stated otherwise. Differences between groups were assessed using Fisher's exact probability test for assessment of equality in proportions for categorical variables, and Mann–Whitney *U* test for univariate comparisons between non-cardiac ATTR patients and controls, as well as between cardiac ATTR and HCM patients. A p value $<.05$ was considered statistically significant. For assessment of RV global and RV FW strain reproducibility, repeated measurements were performed in 10 randomly selected patients by two experienced investigators. Intra- and inter-observer variability was calculated and variability was expressed as the coefficient of variation (standard deviation of the differences between the two sets of measures, divided by the overall mean).

Results

Demographics

The final study population comprised of 42 cardiac-ATTR patients, 19 non-cardiac ATTR patients, 25 HCM patients and 30 healthy adult controls. Demographics of the four groups are outlined in Table 1. Non-cardiac ATTR-patients were significantly younger than controls ($p = .005$), as were HCM with respect to cardiac ATTR patients ($p = .0001$).

HCM patients also had significantly higher median weight than cardiac ATTR patients ($p = .028$).

Left ventricular geometry and function

Apart from a significantly higher heart rate ($p < .0001$), non-cardiac ATTR patients did not differ from healthy controls in any LV geometrical or function parameters (Table 2). HCM patients had significantly thicker IVST ($p = .017$) but thinner PWT ($p = .004$), and thus greater asymmetrical wall thickening (IVST/PWT, $p < .0001$), compared to cardiac ATTR patients. HCM patients also had higher E/A-ratio ($p = .028$). All patients had normal extent of pericardial effusion.

Right ventricular geometry and function

RVT was measured in 68% of the non-cardiac ATTR patients, 81% of cardiac ATTR, 60% of HCM and 93% of controls. Only one patient with non-cardiac ATTR had slightly increased RVT (6 mm) compared with 57% of cardiac ATTR patients and 35% of HCM patients. While RV systolic function, estimated by TAPSE, was preserved in all non-cardiac ATTR patients, it was reduced in 24% with cardiac ATTR and 8% of HCM patients; these proportions did not significantly differ between the two latter patient groups.

There was no difference in RVEDA, RVFAC or RV-RA peak systolic gradient on comparison of cardiac ATTR and HCM groups. In the groups as a whole, there was no difference in RV structure or function between healthy controls and non-cardiac ATTR. Tissue Doppler recordings of the RV free wall were lacking in most HCM patients and could thus not be analyzed (Table 2). We found a positive correlation on comparison of RVEDA and left ventricular end diastolic volume (LVEDV) in both cardiac and non-cardiac ATTR ($r=0.45$, $p=.006$ and $r=0.64$, $p=.006$), however, this difference was not seen in HCM patients.

RV strain analysis

RV segmental and global strain values for all four groups are shown in Table 3. Successful RV endocardial tracings were achieved in 68%, 62%, 60% and 71% of non-cardiac ATTR, cardiac ATTR, HCM and controls, respectively.

The main reasons for unsuccessful RV strain analysis were poor visualization of the RV free wall and RV free wall movement out of the image sector. Patients with cardiac ATTR and HCM exhibited impaired RV global strain ($p=.005$ and $p=.014$, respectively) and RV FW strain ($p=.001$ and $p=.002$, respectively) on comparison to healthy controls. While RV global and FW strain did not differentiate cardiac ATTR from HCM, segmental RV strain was different between the two groups, being lower in the basal free wall region in cardiac ATTR patients ($p=.016$), however, lower in the apical region in the HCM patients ($p=.014$), Figure 1. Consequently, a positive RV apex-to-base strain gradient with relative apical sparing was shown in cardiac ATTR patients, which was significantly different from the pattern seen in HCM patients ($p=.002$). Segmental RV strain patterns in the four study groups are shown in Figure 2. We found no correlation between RV and LV apical sparing in either cardiac ATTR or in HCM patients.

Table 3. Left and right ventricular longitudinal peak strain in patient groups and controls.

Peak systolic strain, %	Cardiac ATTR (n = 42)	HCM (n = 25)	p Value*	Non-cardiac ATTR (n = 19)	Controls (n = 30)	p Value†
LV global	-16.7 (-13.8 to -19.4)	-15.9 (-12.1 to -17.8)	.256	-19.6 (-15.2 to -22.3)	-18.3 (17.7 to -21.9)	.920
RV basal FW	-14.9 (-11.5 to -28.5)	-27.5 (-18.3 to -37.7)	.016	-32.9 (-21.9 to -47.5)	-33.8 (-27.6 to -44.1)	.627
RV mid FW	-18.4 (-13.6 to -28.9)	-18.1 (-11.4 to -32.6)	.893	-22.1 (-17.8 to -30.7)	-27.6 (-15.4 to -35.3)	.578
RV apical FW	-27.3 (-18.5 to -36.1)	-17.5 (-7.6 to -28.7)	.014	-25.8 (-17.6 to -36.0)	-28.6 (-24.1 to -37.7)	.362
RV basal septal	-18.0 (-12.4 to -26.8)	-20.4 (-12.6 to -24.4)	.865	-20.2 (-17.6 to -30.5)	-18.5 (-15.0 to -23.6)	.369
RV mid septal	-16.1 (-12.7 to -19.9)	-15.1 (-10.7 to -17.9)	.360	-18.3 (-14.1 to -28.5)	-16.6 (-10.4 to -23.6)	.276
RV apical septal	-18.9 (-13.0 to -22.6)	-16.1 (-10.3 to -21.2)	.136	-19.4 (-17.3 to -25.4)	-18.3 (-13.6 to -25.2)	.484
RV FW	-23.3 (-16.3 to -26.9)	-23.1 (-17.6 to -26.3)	.981	-28.1 (-24.0 to -29.6)	-30.0 (-25.4 to -33.6)	.170
RV global	-20.3 (-16.7 to -23.6)	-19.7 (-16.1 to -23.5)	.650	-23.1 (-21.5 to -27.1)	-23.3 (-22.2 to 27.4)	.880
RV apical sparing	0.7 (0.5–0.8)	0.4 (0.3–0.6)	.002	0.5 (0.3–0.6)	0.5 (0.3–0.7)	.762

ATTR: transthyretin amyloidosis; FW: free wall; HCM: hypertrophic cardiomyopathy; LV: left ventricular; RV: right ventricular.

*Statistical differences between cardiac ATTR and HCM.

†Statistical differences between non-cardiac ATTR and controls.

Significant differences ($p < .05$) are marked in bold.

Figure 1. Examples of RV peak systolic strain in HCM (B) and ATTR cardiomyopathy (A).

Figure 2. Peak systolic longitudinal strain measured in basal, mid and apical regions of the right ventricle in patients with cardiac and non-cardiac transthyretin (ATTR) amyloidosis, hypertrophic cardiomyopathy (HCM), and healthy controls.

Reproducibility

Intra-observer variability was 14% and 13% for global RV strain and RV FW strain, respectively. Inter-observer variability was 11% for global RV strain and 13% for RV FW strain.

Discussion

The main findings of this study were that; RV involvement is frequently encountered in Swedish ATTR cardiomyopathy patients with concurrent increased LV wall thickness, and that two distinct regional patterns of RV strain were displayed, one for cardiac ATTR and another for HCM patients.

HCM, irrespective of its phenotypic presentation, is characterized by hypertrophied myocytes and myocardial fiber disarray [24]. Comparatively, ATTR amyloidosis is characterized by segmental amyloid infiltration of the myocardium with resulting amyloid deposits that increase wall thickness [25–27]. Thus, LV phenotypic discrimination of the two conditions is achievable. In contrast to this, RV involvement in the two conditions presence, to some extent, similar in terms of prevalence of increased wall thickness and reduced TAPSE, a universally used marker of RV systolic function. In addition, no difference was found for RVFAC or estimated pulmonary pressures, which might have explained the difference in RV apical sparing. Furthermore, intrinsic RV myocardial function was globally reduced in the both conditions; however, the only discriminating variable was the regional strain function, which was reduced in RV apical segments in HCM, but in basal segments in the cardiac ATTR group. Preserved RV apical strain in cardiac ATTR patients is similar to previous descriptions of the LV, in which relative apical sparing has been shown useful for differentiating cardiac amyloidosis from other causes of increased LV wall thickness [23,28,29]. This unique finding, regarding the segmental RV strain pattern in cardiac ATTR, raises a number of questions

that need answers in order to characterize and understand further the RV's involvement in cardiac ATTR.

Since none of our patients had isolated apical HCM, we take this finding as potential subclinical involvement of RV apex with the HCM pathology. A similar loss of RV strain from base to apex in patients with HCM disease was also reported by Badran et al. [30]. In contrast, cardiac amyloidosis seems to initially affect the basal parts of the LV, as seen by lower strain values in the basal segments [31,32].

A potential mechanism behind the difference in apical RV strain in the two diseases could be due to the difference in systolic apical rotation. Apical counter-clockwise systolic strain is reportedly higher in HCM patients compared to amyloidosis patients [33,34]. The increased apical motion in HCM may increase wall stress in the apical segment of the RV and reduced strain.

The present study did not uncover any early markers of RV dysfunction in non-cardiac ATTR patients. Apart from increased RV free wall thickness in one patient, non-cardiac ATTR displayed similar RV size and function as seen in healthy controls. This is in contrast to previous findings in AL amyloidosis, where early impairment of RV longitudinal systolic function, marked by reduced RV free wall strain and TAPSE, was demonstrated in patients despite having normal LV size, thickness and function [11]. These findings are not surprising and are in line with the general understandings that ATTR amyloidosis patients have a more benign clinical course, compared to the rapid disease progression usually occurring in AL amyloidosis [35]. Furthermore, the non-cardiac ATTR patients were relatively young, predominantly had early onset of disease (<50 years of age), and presented mainly with autonomic neuropathy and conduction disturbances clinically [36].

It could be argued that impaired RV function in cardiac ATTR patients is mainly a result of LV systolic or diastolic dysfunction, with backward failure as direct consequence of severe left sided heart failure with elevated left side filling pressures. Naturally, some degree of RV dysfunction in both HCM and ATTR patients could be attributed to the thickened IVST, as the RV and LV share the IVST. However, E/A ratio and E/e' reported in this study present no evidence for severely increased LV filling pressures. In addition, myocardial wall thickness was not severely increased in cardiac ATTR patients indicating that the patients were not in severe dysfunction. Therefore, it seems more likely that the reduction in basal RV free wall strain is predominantly due to amyloid infiltration and deposition, and this hypothesis is supported by an increased RV wall thickness. Furthermore, previous histological studies have shown presence of amyloid deposition also in the right heart [37].

Limitations

As this study was performed retrospectively, specific image acquisition focusing on the RV was not obtained in all patients, which affected the feasibility for RV strain tracings. The interquartile range for RV strain was relatively wide in this study. However, as the intra- and inter-observer variability for RV global and RVFW strain was acceptable, this

variability might be explained by variable stages of disease within the patient groups. Age differences were found in all patients, including between ATTR cardiomyopathy and HCM patients. We had no data on different biomarkers in patients, which would be of interest. Lastly, no ATTR patient underwent endomyocardial biopsy, thus we chose to define non-cardiac amyloidosis based by the echocardiographic criterion of septal thickness >12 mm. Minor amyloid deposits might therefore be present in the non-cardiac ATTR group and undetectable by echocardiography.

Conclusions

Right heart involvement is common in ATTR amyloidosis patients with concomitant increase in LV wall thickness. This study presents an apex-to-base RV strain gradient in ATTR cardiomyopathy that discriminates ATTR from HCM. This RV apical sparing pattern in ATTR is a mirror of the strain pattern previously described for the LV. Further studies are needed to confirm these findings.

Disclosure statement

No potential conflict of interest was reported by the authors.

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