

### P2991 | BEDSIDE

#### An ECG/ECHO comparison between AL and ATTR cardiac amyloidosis at diagnosis

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**Purpose:** Despite a similar pattern of concentric hypertrophy and diastolic dysfunction, a lower prevalence of low-voltage ECG has been reported in hereditary transthyretin-related (mATTR) when compared with light-chain (AL) cardiac amyloidosis. A detailed comparison of all ECG-derived variables is still lacking.

**Methods:** At diagnosis, 12-lead ECG and echocardiography were recorded in consecutive cardiac AL (n=141) or mATTR (n=57) patients. PQ, QRS, QT and heart rate corrected QT intervals, as well as the prevalence of atrio-ventricular (AV) and intra-ventricular (IV) conduction delays, fragmented QRS (fQRS), low QRS voltages and pseudonecrosis were evaluated. QRS score was obtained as the sum of the Q, R and S wave amplitude in all leads.

**Results:** Despite higher LV mass index (an indirect measure of cardiac amyloid deposition), mATTR CA is characterised by higher QRS voltages, as assessed by either QRS score or the prevalence of a low-voltage pattern, by a comparable percentage of pseudonecrosis, by a lower prevalence of fQRS, and by higher prevalence of IV (but not of AV) conduction defects. Moreover, corrected QT interval was shorter and strain-like repolarisation abnormalities were less frequent in mATTR patients, whose heart rate was comparable to AL patients.

Table 1

	AL	mATTR	P-value
Heart Rate (bpm)	78±13	74±11	NS
PQ (msec)	173±34	192±38	0.001
QRS (msec)	90±2	103±28	<0.001
QT corrected	457±39	438±29	0.001
QRS score (mV)	87±41	104±29	0.005
LVMI (g/m <sup>2</sup> )	161±51	213±59	<0.001
Low-voltages pattern (%)	67	31.5	<0.0001
IV conduction delays (%)	26	43.8	0.025
Strain-like repolarization (%)	37.8	10.5	<0.0001

Values expressed as mean ± SD.

**Conclusions:** Despite a higher amyloid "load"/deposition, mATTR is characterised not only by higher QRS voltages, but also by a lower prevalence of repolarisation abnormalities and a higher presence of IV conduction disturbances. These findings, associated with the known worse survival of cardiac AL vs. mATTR, may suggest higher cardiac toxicity of AL vs mATTR amyloid fibrils.

### P2992 | BEDSIDE

#### Dual-chamber pacing and alcohol septal ablation in hypertrophic obstructive cardiomyopathy - results of long-term follow-up

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**Background:** Hypertrophic obstructive cardiomyopathy patients with drug-refractory symptoms could be treated non-pharmacologically by alcohol septal ablation (ASA) and dual-chamber (DDD) pacing. ASA is the most frequently used intervention; DDD pacing is currently used rather exceptionally. However, data from long-term follow up of pacing treated patients show a more pronounced effect than described previously.

**Aim of the study:** To compare the long-term effect of DDD pacing and ASA in the treatment of obstructive hypertrophic cardiomyopathy (HCM).

**Methods:** There were included 24 HCM patients treated with DDD pacing and 52 treated with ASA, who were followed for 101±49 and 87±23 months, respectively. Changes in echocardiographic parameters and functional status of the patients were compared.

**Results:** In the group treated with DDD pacing the left ventricle outflow tract gradient (LVOTG) decreased from 82±44 mmHg to 21±21 mmHg, and NYHA class improved from 2.7±0.5 to 2.1±0.6 (both p < 0.001). In the ASA-treated group a decline in LVOTG from 73±38 mmHg to 24±26 mmHg and reduction in NYHA class from 2.8±0.5 to 1.7±0.8 (both p < 0.001) was observed. The LVOTG change was similar in both groups (p = 0.260), symptoms were more affected by ASA (p = 0.001).

**Conclusion:** ASA and DDD pacing were similarly effective in reducing LVOTG. The symptoms improvement was more pronounced in patients treated with ASA.

### P2993 | BEDSIDE

#### Prospective screening of Afro-Caribbean heart failure patients to determine the prevalence of cardiac transthyretin (ATTR) amyloidosis

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**Purpose:** 4% of African Americans possess the transthyretin (TTR) V122I polymorphism, which is associated with late onset cardiac amyloidosis. However, the incidence of clinically significant disease is not known and the diagnosis is readily missed due to clues on echocardiography being attributed to hypertensive heart disease, along with the misconception that the ECG always demonstrates low voltage complexes in amyloidosis. We previously demonstrated that 10% of Afro-Caribbean patients attending a heart failure clinic in South London have ATTR V122I cardiac amyloidosis, and we report here a simple diagnostic algorithm that we applied to an Afro-Caribbean patient population at another large tertiary cardiac centre in London.

**Methods:** We prospectively reviewed the echocardiograms of Afro-Caribbean patients with non-ischaemic cardiomyopathy over a 6 month period. Patients with increased ventricular wall thickness (> 12 mm) and no clinical diagnosis of hypertrophic cardiomyopathy were referred for cardiovascular magnetic resonance (CMR) and 99mTc-DPD scintigraphy. Patients in whom cardiac imaging suggested amyloidosis were further investigated at the National Amyloidosis Centre.

**Results:** 49 Afro-Caribbean patients presented during the screening period, 11 of whom (22%) met the criteria for further investigation (median age 67 (55-77); 72% males). DPD scintigraphy was performed in all 11 patients and CMR in the 6 patients without a contraindication. A diagnosis of amyloidosis was made in 3 patients (27% of those referred for specialist review and 6% of the total number screened), two of whom had the TTR V122I variant and one had wild-type ATTR amyloidosis.

**Conclusions:** Cardiac amyloidosis is an important cause of heart failure in the elderly Afro-Caribbean population and the diagnosis should be considered when echocardiography suggests "LVH". DPD and CMR imaging are valuable diagnostic tools. Several specific therapies for ATTR amyloidosis are currently in clinical development, underscoring the need for accurate diagnosis.

### P2994 | BEDSIDE

#### Cardiac magnetic resonance: evaluation of signal intensity decay after the injection of gadolinium in cardiac amyloidosis

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**Purpose:** Late Gadolinium Enhancement (LGE) technique is currently used for the diagnosis of Cardiac Amyloidosis (CA) by Magnetic Resonance (MRI). However, LGE pattern lacks of sensitivity because it is absent in 30% of patients with CA in previous series. Aim: to test a new method for diagnosis of CA by the evaluation of curves of myocardial signal intensity (SI)/time after gadolinium injection.

**Methods:** Cardiac MRI examination was performed in 60 consecutive patients (males 37 [62%], mean age: 69±10 years) with previous diagnosis of cardiac AL amyloidosis and 10 healthy control subjects (6 males [60%], mean age: 65±14 years). Protocol included evaluation of morphologic and functional features of CA. Conventional LGE images were also acquired and the presence, extent and pattern of LGE was evaluated. Acquisition of T1-weighted inversion recovery gradient echo horizontal long axis view (inversion time, 250 msec) was repeated every minute after gadolinium injection for 8 minutes. In all the images regions of interest (ROIs) were placed in the following sites: subendocardium, subepicardium, middle layer of interventricular septum, left ventricular cavity and skeletal muscle (the latter considered as reference ROI). A software automatically measured SI for all the ROIs generating a SI/time curve, where time was expressed in heart beat number. For each ROI half time of MRI-signal decay (T1/2) was calculated from the SI/time curves.

**Results:** The typical LGE pattern for CA was observed in 43/60 patients (72%), while 17 subjects showed morphologic features of CA (indexed left ventricular mass 92.7±34.4 g/m<sup>2</sup>), but negative LGE. At ROC analysis subendocardial T1/2 allowed to distinguish patients with typical LGE from controls with a sensitivity of 98% and specificity 89% (threshold value >272 beats; area under the curve 0.95). Using the same threshold (>272 beats), an abnormal SI/time curves was detected in 12/17 patients with CA and negative LGE. Then, the analysis of SI/time curve permitted to evidence CA in 92% of patients.

**Conclusions:** The analysis of SI/time curve after gadolinium injection, which represents a simple way to evaluate gadolinium kinetics in myocardium and blood, may improve the accuracy of MRI-LGE for the diagnosis of CA.