Preliminary evaluation of stress myocardial blood flow pattern in patients with apical hypertrophic cardiomyopathy candidate to be enrolled in the CARAPACE study.

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Funding Acknowledgements: Net-2011-02347173 Italian Ministry of Health

Background. Microvascular dysfunction (MVD) plays a major role in the pathophysiology of hypertrophic cardiomyopathy (HCM). So far there is no clear demonstration that medical therapy can favorably affect HCM through an improvement in MVD. The CARAPACE study financed by the Italian Ministry of Health is specifically focused on assessing the effect of therapy on global myocardial blood flow (MBF) in HCM.

Aim. During patient screening for potential enrolment in the CARAPACE trial, we encountered a high proportion (30%) of subjects with the apical pattern of HCM. Because scanty data are available about MBF behavior in these patients, we aimed to assess their stress MBF pattern, and to compare it with that of "classic" septal HCM, in order to exclude a potential selection bias in the CARAPACE cohort.

Methods. In our database we enrolled 12 patients, 10 males and 2 females (median age 44, interquartile range 26 - 60) with confirmed diagnosis of apical HCM, referred for stress quantitative 13NH3positron emission tomography (PET) in our Nuclear Medicine Department. Regional perfusion analysis, stressMBF, and stress transmural perfusion gradient (TPG = subendocardial / subepicardial MBF) were assessed using the Pcard tool of the Pmod processing platform. Data were compared with our historical PET database of HCM patients (n = 100).

Results. Regional perfusion analysis visually demonstrated apical hypoperfusion in 10 of 12 patients (83%), with wide perfusion defect in 3 patients. Taking as reference our recently published prognostic classification in global stress MBF tertiles, flow values were in the highest tertile (≥2.14 mL/min/g) for 3 patients, in the middle tertile (1.54 to 2.13 mL/min/g) for 5 patients, and in the lowest tertile (≤1.53 mL/min/g) for 4 patients. The apical stress TPG was abnormal (<1) in 5 patients and normal in the remaining 7. There was no correlation between the global stress MBF and abnormal apical stress TPG.

Conclusion. The apical variant of HCM is associated with regional stress hypoperfusion of the apex in the majority of patients. Similar to septal HCM, global stress MBF values are heterogeneous and distributed within the whole disease-specific spectrum of flow-values, with no correlation between global MBF values and visual detection of perfusion defects. There was no correlation between global stress MBF values and visual detection of perfusion defects. There was no correlation between global stress MBF values and abnormal apical TPG. Collectively, these data are in agreement with those observed in the classical septal HCM, despite the peculiar anatomy and genetic basis. These data support the inclusion of apical HCM in the CARAPACE trial.