Clinical spectrum and outcome of advanced heart failure in hypertrophic cardiomyopathy: from pathophysiology to contemporary unmet needs

Authors:
G Galati1, S Zieroth2, A Di Lenarda3, N Aspromonte4, I Vranic5, M Gulizia6, A Margonato1, 1San Raffaele Hospital and Scientific Institute (IRCCS), Heart Failure Unit and Division of Cardiology, Cardiothoracic and vascular Department - Milan - Italy, 2St. Boniface General Hospital, Heart Failure and Transplant Clinics, Section of Cardiology - Winnipeg - Canada, 3Cardiovascular Center A.S.S. 1 of Trieste, Cardiovascular Department - Trieste - Italy, 4Polyclinic Agostino Gemelli, Cardiology Unit - Rome - Italy, 5Clinical Centre of Serbia, Cardiology Department - Belgrade - Serbia, 6Garibaldi Hospital, Cardiology Unit - Catania - Italy,

On behalf: Heart Failure in Hypertrophic Cardiomyopathy

Topic(s):
Myocardial Disease – Clinical

Citation:

Background

Advanced heart failure (AdvHF) in hypertrophic cardiomyopathy (HCM) was overlooked. Neither large case series nor clinical trials on this topic have been described. The main clinical-pathological profiles responsible for AdvHF in HCM are: 1) End-stage HCM (ES-HCM) defined by an ejection fraction (EF) =50%; 2) LV outflow obstruction despite optimal pharmacological and not pharmacological therapy (Refractory HOCM); 3) Nonobstructive HCM with preserved EF (HNOCMpEF).

Purpose

Based on a systematic revision of all published manuscripts on this topic, this study describes prevalence and outcomes of the 3 main HCM phenotypes responsible for AdvHF, heart transplantation (HTx), left ventricular assist device (LVAD) implantation and death for heart failure (HF-death) with the contemporary management of HCM.

Methods

The study screened 120 manuscripts in MEDLINE and EMBASE on AdvHF in HCM patients published from 2000 until December 2018, in adult patients (=18 years old). The authors identified 8 manuscripts eligible for the analysis. 205 patients with AdvHF due to HCM, despite optimal therapy, were included in the main analysis. AdvHF was defined in presence of severe NYHA symptoms (class III and IV), because in all the manuscripts this definition was used. Mean follow-up = 7.6 years.

Results

Figure 1 shows the prevalence of phenotypes responsible for AdvHF and HTx/LVAD implantation/HF-Death. Please notice on the right the combined outcome split in two HTx/LVAD implantation separated from HF-Death. Of 205 HCM patients, 119 (58%) underwent HTx, LVAD implantation or died for HF. Figure 2 shows the outcome per each clinical phenotype.

Conclusion

AdvHF in HCM has an ominous prognosis, indeed 58% of patients underwent HTx/LVAD implantation or died
for HF. AdvHF in HOCM has a good outcome with the contemporary management. Less than 1/3 of cases of AdvHF in HCM was determined by HNOCMpEF due to massive hypertrophy and restrictive physiology and only 16.4% of them died due to HF. Nowadays, ES-HCM represents the main cause of AdvHF in HCM (64.9% of all HCM patients) and the major determinant for poor outcome (74.8% of Htx/LVAD/HF-death among all HCM patients). Although it has been managed with HTx or LVAD implantation, 1/3 of these patients died due to HF. This reflects poor attention and portrays an unmet need for HCM patients, in particular for ES-HCM patients that are younger than all others HCM patients. These findings reinforce the emphasis on long-term surveillance of HCM patient in order to timely identify patients at risk of ES evolution and early start the standard HF therapeutic (pharmacological and non-pharmacological) armamentarium. Finally, developing specific therapies for these HCM patients it is of paramount importance.