If the result for a patient is more than 12.78 then the risk of congestive HF is 10 times higher with relative risk (RR) 10.0 (95% CI 3–33.3) with specificity - 81% and sensitivity - 79%, OR 15.5 (95% CI 3.9–61.3, p=0.0001).
Conclusion: The proposed model enables one to predict congestive HF development in HCM patients on the base of LA diameter, LWPW thickness, LVDD degree and presence of permanent AF.

P2597
Clinicopathological profiles responsible for advanced heart failure, heart transplantation, left ventricular assist device implantation and death for heart failure in Hypertrophic cardiomyopathy

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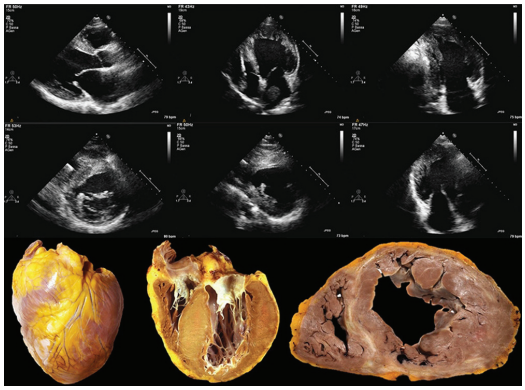
Background: Over the last 50 years, advanced heart failure (AdvHF) in hypertrophic cardiomyopathy (HCM) was overlooked. Neither large case series nor clinical trials on this topic have been reported. The main clinical-pathological profiles responsible for AdvHF in HCM are: 1) End-stage HCM (ES-HCM) defined by an ejection fraction (EF) ≤50%; 2) Left-Ventricular 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 the prevalence of the three 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 HCM cases of AdvHF and HTx published since 2000 until December 2017, in adult patients (≥18 years old). The authors identified 8 manuscripts eligible for the analysis, 4 of whom were excluded for incomplete information before HTx. 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. Minimum reported follow-up was 6.1 years.
Results: Table 1 shows the prevalence of phenotypes responsible for AdvHF/HTx/LVAD implantation/HF-Death. Of 205 HCM patients, 119 (58%) underwent HTx, LVAD implantation or died for HF.

Table 1

	Adv HF (=205)	HTx/LVAD Implantation/ HF-Death (=119)	HTx/LVAD Implantation (= 68)	HF-Death (=51)
ES-HCM	133 (64.9%)	89 (74.8%)	49 (72.1%)	40 (78.5%)
Refractory HOCM	11 (5.4%)	1 (0.8%)	0 (0%)	1 (1.9%)
HNOCMpEF	61 (29.7%)	29 (24.4%)	19 (27.9%)	10 (19.6%)



Morphological features of End-Stage HCM

Conclusion: AdvHF in HCM has a poor prognosis. With the current management of HOCM only a very little percentage of these patients experiences AdvHF. Less than 1/3 of HNOCMpEF patients had AdvHF due to restrictive physiology and less than 1/4 of these patients had a poor prognosis. Nowadays, ES-HCM represents the main cause of AdvHF in HCM and the major determinant for poor outcome. Although it has been managed with HTx or LVAD implantation, a significant percentage (78.5%) died for HF. This reflects poor attention and portrays an unmet need for ES-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 not) armamentarium

P2598
Long-term outcome in patients with sarcomere gene mutation among Japanese hypertrophic cardiomyopathy populations

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Background: Hypertrophic cardiomyopathy (HCM) is the most common hereditary cardiomyopathy with an autosomal-dominant trait mainly caused by mutation in sarcomere genes. According to the recent studies, sarcomere gene mutations are identified among 40–60% of HCM patients, and family screening of gene mutation is recommended by several guidelines. However, few data exist regarding the long-term clinical impact on presence of sarcomere mutations.
Purpose: We sought to clarify the relationship between presence of these mutations and long-term outcome in HCM.
Method: We studied consecutive 211 Japanese patients with HCM (75 females) who had genetic testing between 2003 and 2013. All patients were diagnosed HCM based on echocardiographic demonstration of unexplained left ventricular (LV) hypertrophy in the absence of systemic hypertension or other cardiac disease (valvular heart disease or storage disease). Demographic, laboratory, echocardiographic data, as well as survival data, were collected from the patients' medical records. Genetic analyses were performed by using direct DNA sequencing for mutations in translated exons of the six common sarcomere genes (MYH7, MYBPC3, TNNT2, TNNI3, TPM1, ACTC). Composite outcome was determined by arrhythmic events (sudden cardiac deaths, successful recovery from sustained ventricular tachycardia or ventricular fibrillation, and appropriate ICD discharge), heart failure events (heart failure deaths, heart failure hospitalizations, and worsening NYHA required additional medication), and embolic events (embolic deaths and embolic hospitalizations).
Results: Overall, 75 (36%) patients were genotype positive for a putative HCM-associated mutation. Patients were divided into two groups by presence of mutations: the mutation positive group and mutation negative group. At baseline, the mutation positive group included younger (57±17 vs. 64±13, p=0.002) and more frequent family history (60 vs. 11%, p<0.0001) patients than the mutation negative group, but there were no significant differences in echocardiographic parameters including maximum left ventricular wall thickness (20±4 vs. 20±4mm, p=0.84), LA diameter (45±8 vs. 44±7mm, p=0.26), and ratio of LV outflow tract obstruction (13.3 vs. 17.7%, p=0.44). Follow-up period were up to 14.5 years, and mean follow-up period was 7.3±4.5years. Survival analysis revealed that the positive gene mutation group had significant worse composite outcome compared with the mutation negative group (log-rank p=0.042, Figure). This tendency was more prominent regarding arrhythmic events (log-rank p=0.002, Figure).

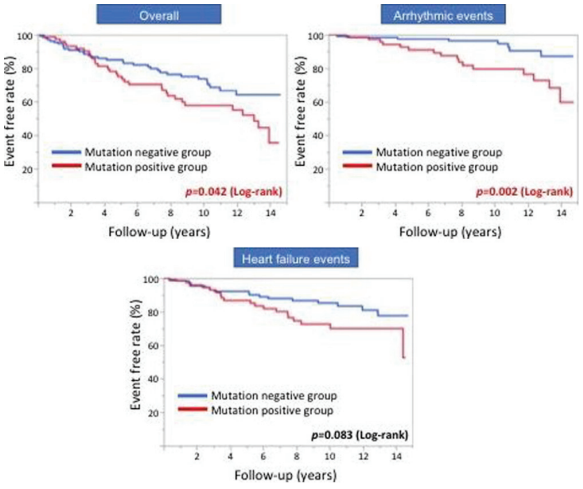

Figure 1

Conclusions: In our HCM cohort, patients with sarcomere gene mutation are associated with poorer long-term outcome than those without mutation. These results suggested that presence of sarcomere gene mutation seem to be one of the useful predictors for HCM-related morbid events, and the genetic information is considered to be important to provide us better management of HCM.

CONGENITAL HEART DISEASE

P2599
The Notch pathway regulatory protein MIB1 is a novel gene for nonsyndromic bicuspid aortic valve

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