Working Group
Heart Failure
Clinical Scenarios

2nd INTERNATIONAL CONGRESS
ON CARDIOVASCULAR IMAGING
IN CLINICAL PRACTICE

Iakovos Armenis
Onassis Cardiac Surgery Center
Conflict of interest: none.
Case

• 70 year old patient with HOCM
• Gradually worsening dyspnoea during the last 3 months (NYHA III)
• 10 years ago: ischemic stroke, moderate aortic stenosis, mean pressure gradient 31mmHg
• 9 years ago: syncope, slightly increased troponin, carotid atheromatosis (<40%)
• AF
Case

• Recent hospitalization: severe LVOT obstruction (pressure gradient 70-125mmHg), mitral valve stenosis (MVA 1,3cm$^2$), mild-moderate aortic valve stenosis
• Coronary angiography: LM 40%, LAD 70%, LCX, RCA without critical stenoses
• Metoprolol 100mg 1x2, atorvastatin 20mg 1x1, acenocumarol 4mg
2016 Rest

Exercise LVO gradient

2016 Bike 25 watts 3 min

Exercise AV gradient
Prevalence and Impact of Coexistent Bicuspid Aortic Valve in Hypertrophic Cardiomyopathy.

Padang R¹, Gersh BJ¹, Ommen SR¹, Geske JB².

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Abstract

BACKGROUND: The association between bicuspid aortic valve (BAV) and hypertrophic cardiomyopathy (HCM) has been reported but its true prevalence is unknown. This study investigated the prevalence and clinical impact of coexistent BAV in a large referral HCM population.

METHODS: Retrospective analysis of 3765 echocardiograms between 2004 and 2014 in 2640 consecutive patients with HCM was performed to assess for BAV. Patients with coexistent conditions were studied.

RESULTS: Twenty-three patients (0.9%) were identified with coexisting BAV and HCM. Mean age was 52±16 years, 18 males (78%), 16 with NYHA functional class I/II at initial evaluation (70%). A family history of HCM was present in five patients (22%); none had a family history of BAV or aortopathy. Maximal left ventricular wall thickness was 24±6 mm; the majority had either reverse curve or sigmoid septal morphology. Moderate or greater aortic valve dysfunction was present in seven patients (30%), BAV-related aortopathy in 18 patients (78%) and dynamic left ventricular outflow tract (LVOT) obstruction in nine patients (39%). Three patients had combined LVOT obstruction and aortic stenosis. Median time from diagnosis of BAV or HCM to last follow-up was 11±12.5 years. At last follow-up, 22% had undergone BAV-related surgeries, 30% had septal reduction therapy (SRT), and 17% had combined SRT and BAV-related surgeries. Overall survival was 95% at 10 years.

CONCLUSIONS: This study reported a 0.9% prevalence of BAV among HCM population, similar to the general population. Aortopathy and LVOT obstruction were common, necessitating cardiac interventions in over one-third of cases. Long-term survival appeared favourable.
Risk stratification and outcome of patients with hypertrophic cardiomyopathy >=60 years of age.

Maron BJ, Rowin EJ, Casey SA, Haas TS, Chan RH, Udelson JE, Garberich RF, Lesser JR, Appelbaum E, Manning WJ, Maron MS.

Abstract

BACKGROUND: Hypertrophic cardiomyopathy (HCM) is prominently associated with risk for sudden death and disease progression, largely in young patients. Whether patients of more advanced age harbor similar risks is unresolved, often creating clinical dilemmas, particularly in decisions for primary prevention of sudden death with implantable defibrillators.

METHODS AND RESULTS: We studied 428 consecutive HCM patients presenting at ≥60 years of age and followed for 5.8±4.8 years; 53% were women. Of the 428 patients, 279 (65%) survived to 73±7 years of age (range, 61-96 years), most (n=245, 88%) with no/mild symptoms, including 135 with ≥1 conventional sudden death risk factors and 50 (37%) with late gadolinium enhancement. Over follow-up, 149 (35%) died at 80±8 years of age, mostly from non-HCM-related causes (n=133, 31%), including a substantial proportion from noncardiac disease (n=54). Sixteen patients (3.7%) had HCM-related mortality events (0.64%/y), including embolic stroke (n=6), progressive heart failure or transplantation (n=3), postoperative complications (n=2), and arrhythmic sudden death events (n=5, 1.2% [0.20%/y]). All-cause mortality was increased in HCM patients ≥60 years of age compared with an age-matched US general population, predominantly as a result of non-HCM-related diseases (P<0.001; standard mortality ratio, 1.5).

CONCLUSIONS: HCM patients surviving into the seventh decade of life are at low risk for disease-related morbidity/mortality, including sudden death, even with conventional risk factors. These data do not support aggressive prophylactic defibrillator implantation at advanced ages in HCM. Other cardiac or noncardiac comorbidities have a greater impact on survival than HCM in older patients.
Hypertrophic Cardiomyopathy in Adulthood Associated With Low Cardiovascular Mortality With Contemporary Management Strategies.

Maron BJ1, Rowin EJ2, Casey SA3, Link MS2, Lesser JR3, Chan RH2, Garberich RF3, Udelson JE2, Maron MS2.

Abstract

BACKGROUND: Hypertrophic cardiomyopathy (HCM) has been prominently associated with adverse disease complications, including sudden death or heart failure death and a generally adverse prognosis, with annual mortality rates of up to 6%.

OBJECTIVES: This study determined whether recent advances in management strategy, including implantable cardioverter-defibrillators (ICDs), heart transplantation, or other therapeutic measures have significantly improved survival and the clinical course of adult HCM patients.

METHODS: We addressed long-term outcomes in 1,000 consecutive adult HCM patients presenting at 30 to 59 years of age (mean 45±8 years) over 7.2±5.2 years of follow-up.

RESULTS: Of 1,000 patients, 918 (92%) survived to 53±9.2 years of age (range 32 to 80 years) with 91% experiencing no or only mild symptoms at last evaluation. HCM-related death occurred in 40 patients (4% [0.53%/year]) at 50±10 years from the following events: progressive heart failure (n=17); arrhythmic sudden death (SD) (n=17); and embolic stroke (n=2). In contrast, 56 other high-risk patients (5.6%) survived life-threatening events, most commonly with ICD interventions for ventricular tachyarrhythmias (n=33) or heart transplantation for advanced heart failure (n=18 [0.79%/year]). SD occurred in patients who declined ICD recommendations, had evaluations before application of prophylactic ICDs to HCM, or were without conventional risk factors. The 5- and 10-year survival rates (confined to HCM deaths) were 98% and 94%, respectively, not different from the expected all-cause mortality in the general U.S. population (p=0.25). Multivariate independent predictors of adverse outcome were younger age at diagnosis, female sex, and increased left atrial dimension.

CONCLUSIONS: In a large longitudinally assessed adult HCM cohort, we have demonstrated that contemporary management strategies and treatment interventions, including ICDs for SD prevention, have significantly altered the clinical course, now resulting in a low disease-related mortality rate of 0.5%/year and an opportunity for extended longevity.