Treatment of Hypertrophic Cardiomyopathy in 2017

Bruce B. Reid, MD
Disclosures

• I have no conflicts of interest to disclose

• I will not be discussing any off label medications and/or devices
Objectives

• 1) Understand the manifestations and treatment for HCM

• 2) Understand the importance of specialized HCM Centers
Hypertrophic Cardiomyopathy

- Myocardial hypertrophy without obvious cause
- Significant cause of sudden cardiac death
- Frequently asymptomatic
- Common: ~ 1 in 500 (0.2% of general population)
- Mutation of one of nine genes
- Results in a mutation of a protein of the sarcomere
- Myocardial disarray
• Typically an autosomal dominant trait
• Most common genetic cardiovascular disorder
• Mutation of genes that encode sarcomere proteins
• Offspring: 50% chance of having the mutation
• Majority involve the B-myosin heavy chain
• Patients without family history: de novo mutation
• Genetic testing available to identify relatives at risk
Two Broad Categories of HCM

1. Obstructive (HOCM, IHSS, ASH)
   - Defined as resting or provokable left ventricular outflow tract gradient > 30mmHg
   - 2/3 of HCM patients are obstructive

2) Non-obstructive – no obstruction at rest or with provocation
Clinical Manifestations of HCM

Symptoms:
- Palpitations
- Syncope
- Dyspnea
- Chest pain
- Sudden death
- Stroke

Causes of symptoms:
- Atrial fibrillation
- Ventricular tachycardia
- LVOT obstruction
- Diastolic dysfunction
- Coronary microvascular dysfunction
Severe HCM
Atrial fibrillation

• Common – about 20-25% of HCM patients
• Diagnose asymptomatic patients with annual Holter monitor
• High risk of stroke/embolic phenomena in HCM patients with atrial fibrillation
• **Always need anticoagulation.** Do not use CHADS2 or CHADS2-vasc score. (consensus recommendations from ACC/AHA/HRS guidelines)
Ventricular Arrhythmias/SCD

- HCM patients have propensity of VT and sudden death (10-15% need ICDs)
- Selective ICD use based on risk stratification
- Highest risk are those with unexplained syncope, family history of early SCD, and severe hypertrophy (> 3.0 cm)
- Novel risk factors using cMRI are starting to be used for assessment
- Annual Holter monitors should be used to assess for asymptomatic VT
Genetic Testing/Family Screening

- HCM is a genetically determined disease: autosomal dominant
- We are aware of 60-70% of the mutations responsible for causing HCM.
- ACC/AHA recommends genetic testing in all HCM patients
- If genetic testing not available or patient declines, primary relatives should be screened with echocardiography and EKG
LVOT Obstruction

- 2/3 of patients with HCM have LVOT obstruction due to systolic anterior motion of the mitral valve (SAM)
- LVOT obstruction worsens with decreased preload, decreased afterload (vasodilation), and increased contractility
- **Need to avoid vasodilators and diuretics** (ACEi/ARB, amlodipine, nitrates, hydralazine, HCTZ, furosemide)
- **Beta-blockers are preferred** (metoprolol, atenolol) to decrease HR and contractility. Goal HR 50-60 at rest. Diltiazem/verapamil can also be used.
LVOT Obstruction

- In symptomatic patients with LVOT gradients above 50mmHg despite maximal medical therapy, septal reduction therapy is recommended.
- Two choices for septal reduction:
  - 1) Surgical myectomy
  - 2) Alcohol septal ablation
- Which therapy to choose is a complex decision--dependent on patient’s anatomy, age, comorbidities.
Alcohol Septal Ablation

- Percutaneous procedure performed in the cath lab: focal septal infarct with the goal of decreasing septal thickness near LVOT
- Generally used in older patients and others that are poor surgical candidates
- Limited by septal perforator anatomy
- 20% of patients develop complete heart block requiring pacemaker
Surgical Myectomy

• Open surgery using cardiopulmonary bypass
• Surgical technique has evolved in specialized centers from a limited myectomy to a more extended resection
• Ideally performed at centers with volumes of 30 cases or more per year (per ACC/AHA guidelines)
Septal Myectomy
Septal Myectomy
Case Presentation
Dynamic LVOT Obstruction
Dynamic LVOT Obstruction
Systolic Anterior Motion of Mitral Valve
Resting Gradient

FR 50Hz
15cm

2D
77%
C 50
P Off
Gen

Vel 440 cm/s
PG 77 mmHg

CW
70%
2.5MHz
WF 225Hz

PAT T: 37.0C
TEE T: 39.1C
75mm/s
80bpm
Septal Myectomy
Postop Gradient

FR 50Hz
16cm
2D
C 50
P Off
Gen

CW
40%
2.5MHz
WF 225Hz

Vmax 138 cm/s
Max PG 8 mmHg
Mean PG 5 mmHg
VTI 18.1 cm
ACC/AHA Guidelines for HCM

ACC/AHA Guideline

2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy

A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines

Developed in Collaboration With the American Association for Thoracic Surgery, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

WRITING COMMITTEE MEMBERS*

Bernard J. Gersh, MB, ChB, DPhil, FACC, FAHA, Co-Chair*†; Barry J. Maron, MD, FACC, Co-Chair*†;
Robert O. Bonow, MD, MACC, FAHA‡; Joseph A. Dearani, MD, FACC§||;
Michael A. Fifer, MD, FACC, FAHA*†; Mark S. Link, MD, FACC, FHRS*¶||;
Srihari S. Naidu, MD, FACC, FSCAI*#; Rick A. Nishimura, MD, FACC, FAHA†;
Steve R. Ommen, MD, FACC, FAHA†; Harry Rakowski, MD, FACC, FASE†**;
Christine E. Seidman, MD, FAHA†; Jeffrey A. Towbin, MD, FACC, FAHA††;
James E. Udelson, MD, FACC, FASNC‡‡§§; Clyde W. Yancy, MD, FACC, FAHA|||
2.2.4. Hypertrophic Cardiomyopathy Centers

The writing committee considers it important to emphasize that HCM is a complex disease entity with a broad (and increasing) clinical and genetic spectrum. Although HCM is one of the most common forms of genetic heart disease and relatively common in the general population, this disease entity is infrequent in general clinical practice, with most cardiologists responsible for the care of only a few patients with HCM. This principle has led to an impetus for establishing clinical programs of excellence—usually within established centers—in which cardiovascular care is focused on the management of HCM (i.e., “HCM centers”). Such programs are staffed by cardiologists and cardiac surgeons familiar with the contemporary management of HCM and offer all diagnostic and treatment options, including genetic testing and counseling, comprehensive transthoracic echocardiogram (TTE), CMR imaging, both surgical septal myectomy and alcohol ablation, and the management of atrial fibrillation (AF)/atrial flutter, and ICDs. Another advantage is the potential to perform outcomes research on large groups of patients. Although the writing committee does not necessarily recommend that all patients with HCM should be evaluated in such centers, nevertheless, it is the strong view that patients with this disease may well benefit from a clinical environment with specific expertise in HCM. The selection of patients for referral to an HCM center should be based largely on the judgment of the managing cardiologist and the degree to which he or she is comfortable advising and evaluating patients with HCM with a particular clinical profile.
HCM Center of Excellence

- Designation by the Hypertrophic Cardiomyopathy Association to select HCM Centers providing comprehensive, contemporary, multidisciplinary care for HCM patients
- ~25 HCM Centers of Excellence (CoE)
- Intermountain Medical Center is the only CoE in Utah and surrounding states (Idaho, Nevada, Wyoming, Arizona, Montana, Colorado)
Initial evaluation:
- Clinic visit with HCM cardiologist
- Echocardiogram
- EKG
- Holter monitor
- Cardiac MRI (in most)
- Genetic counseling/testing (in most)
- +/- Stress test to assess LVOT gradient with exercise
- Referral for septal reduction therapy as necessary
Intermountain Heart Institute

Hypertrophic Cardiomyopathy Center:
801-507-8HCM (8426)