Hypertrophic Cardiomyopathy Protocols

Revised 10/7/2023

- Diagnosis and Treatment
- Imaging
- Exercise
- Cardiac Myosin Inhibitors
Hypertrophic Cardiomyopathy Protocols

Diagnosis and Treatment
Suspected diagnosis or family history of HCM

- Comprehensive H&P with ECG, Echo

  - Family history
    - Yes
      - Consider genetic testing
      - Patient has family variant
        - Screening ECG and Echo per intervals listed
    - No
      - Screening ECG and Echo per intervals listed

  - High level of clinical suspicion
    - No
      - No further testing needed
    - Yes
      - Phenotypically Positive HCM

- Phenotypically Negative
  - Genetic testing
    - Patient has family variant
      - Screening ECG and Echo per intervals listed
    - Patient does not have variant
      - No further testing needed

- Phenotypically Positive HCM
  - See Treatment Protocol
    - Every 1 – 2 years or change in clinical symptoms: clinical assessment, Echo, & Holter (if no ICD present)

  - SCD Risk Assessment (see SCD and Exercise Protocols)
HCM - Treatment

Phenotypical HCM Diagnosed
Preserved LV Function

Symptoms

Institute medical therapy Beta Blocker (Non vasodilating) +/- Verapamil/ Diltiazem
Avoid vasodilators/ high dose diuretics

LVOT Obstruction at rest

No LVOT Obstruction at rest

Stress Echo to evaluate for exercise LVOT Gradient

No gradient with stress

Gradient with stress (>30mmHg)

Evaluate alternative causes for symptoms

Consider Septal Reduction Therapy

Consider Cardiac Myosin Inhibitors (see CMI pathway)

See Invasive Therapies Protocol

Up titrate to symptoms/ HR/ BP

If Resting or Exercise Gradient > 30 mmHg and continued symptoms

Consider Disopyramide

No symptoms

Consider ETT/CPET to evaluate functional capacity
Repeat every 2 – 3 years

Symptoms develop or decrease functional capacity

See Slide 5

See Slide 17

See Slide 9
HCM - Non-Obstructive Symptomatic

Symptoms with no obstruction

Atrial Fibrillation

See AF Pathway

Heart Failure

Systolic Function

LVEF<50%

LVEF>50%

Treat per HF guidelines for HFpEF

NYHA Class 1-2

NYHA Class 3-4

Continue current therapy

LBBB?

Yes CRT

No

Optimize BB, Ca Blockers, consider Ranexa

Yes

Evaluate for revascularization

Treat for CAD (ASA, Statin)

Optimize GDMT

Re-evaluate symptoms

Consider iCD (See Pathway)

Discontinue negative inotropic agents (Verapamil, Diltiazem, Disopyramide)

Evaluate for other causes of reduced EF

ACEI/ARB/ARNI HF BB/MRA/SGLT2i per HF guidelines

NYHA Class 1-2

LBBB?

Refer for evaluation for ASO (Transplant/ VAD)

Persist NYHA Class 3-4

Recurrent ventricular arrhythmias

Evaluated for other causes of reduced EF

Optimize GDMT

Re-evaluate symptoms

Angina

Consider changing Beta blocker to Verapamil (or add Verapamil)

Evaluate for CAD (MPI, MRI, PET, CCTA, Cath)

NYHA Class 3-4

Yes

No

In highly selected patients with apical HCM with severe dyspnea or angina (NYHA class III or class IV) despite maximal medical therapy, and with preserved EF and small LV cavity size (LV end-diastolic volume <50 mL/m² and LV stroke volume <30 mL/m²). Apical myectomy by experienced surgeons at comprehensive centers may be considered to reduce symptoms.
HCM - Management of Atrial Fibrillation

**Documented Atrial Fibrillation**

- Anticoagulation with Vitamin K antagonist or DOAC to an INR of 2.0 – 3.0, regardless of CHA₂DS₂-VASc Score

**Rate Control or Rhythm Control?**

- Rate Control
  - Beta Blocker
  - Calcium Channel Blocker

- Persistent symptoms or poor rate control
  - AV Node Ablation and CIED Implant

**Rhythm Control**

- Anti arrhythmic drug
- Sotalol
- Disopyramide
- Amiodorone
- Dofetilide
- Dronedarone

**Persistent or recurrent AF**

- Endocardial catheter ablation
- Surgical ablation if undergoing open heart surgery/myectomy
Risk Assessment for Sudden Cardiac Death (SCD)

- Prior cardiac arrest, VF, or sustained VT
  - Yes: ICD recommended
  - No: Family history of SCD in 1st degree relative or LV thickness ≥ 30mm or recent unexplained syncope
    - Yes: Other SCD risk modifiers present?
      - Yes: ICD can be useful
      - No: ICD not indicated
    - No: Abnormal BP response
      - Yes: ICD reasonable
        - See “Consider ICD” on slide 8
      - No: Non-sustained VT
        - Yes: ICD not recommended
          - See “EP Consult” on slide 8
        - No: Extensive LGE on CMR (>15%)
          - Yes: ICD can be considered
          - No: ICD not indicated

- EP Consult
  - Yes: See “EP Consult” on slide 8
  - No: ICD not indicated
HCM – ICD-Risk SCD Model

**Primary Prevention**

- **Recommended Assessment**
  - History
  - 2D/ Doppler TTE
  - 48 hour Ambulatory ECG

- **HCM Risk-SCD Variables**
  - Age
  - Family History of SCD
  - Unexplained Syncope
  - LVOT Gradient
  - LV Wall Thickness
  - LA Diameter
  - NSVT
  - Late Gadolinium Enhancement >15%

- **HCM Risk – SCD Score**

**Consider ICD**

- **EP Consult**

**Secondary Prevention**

- Cardiac arrest due to VT or VF
- Spontaneous sustained VT causing syncope or hemodynamic compromise

- **Life expectancy > 1 year**

**ICD recommended**

**Low Risk**

- 5-year risk <4%

- **ICD generally not indicated**

**Intermediate Risk**

- 5-year risk >4-6%

- **ICD may be considered**

**High Risk**

- 5-year risk > 6%

- **ICD should be considered**
HCM - Invasive Therapies

NYHA III, obstruction >50 mmHg at rest or provocation, despite optimal medical therapy, as defined in treatment pathway.

See slide 4

Coronary Angiogram with invasive gradients at rest and provocation, and review of specific anatomy by MRI

Multidisciplinary Clinic Evaluation (IC & CS) to discuss Septal Reduction

Surgical risk, patient preference, anatomy

Low, intermediate surgical risk

Surgical Myectomy

High surgical risk/ patient preference

Alcohol Septal Ablation

TTE follow-up with IC/ CS at 1 month

Subsequent follow-ups with HCM Cardiologists

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Imaging
HCM - Resting TTE

Resting TTE Study

Images*
- Parasternal long axis, with and without color
- M-mode of mitral valve
- A4C, with and without color
- A5C, with and without color
- A3C, with and without color

LVOT Gradient
Continuous Wave Doppler through the LVOT/aortic valve
- Apical 5 and apical 3 chamber views
- At rest and during Valsalva

Mitral Regurgitation Gradient
Continuous Wave Doppler
- Apical 4 and apical 2 chamber views
- At rest and during Valsalva

Maximal LV Wall Thickness
2D measurements of thickest portion of Left Ventricle
- Parasternal and apical views

*Consider using ultrasound enhancing agents in patients with apical HCM to better assess apical hypertrophy and also assess for apical aneurysm.
HCM - Stress TTE

Treadmill Stress TTE

Obtain Baseline Images
- Parasternal long axis, with and without color
- M-mode of mitral valve
- A4C, with and without color
- A5C, with and without color
- A3C, with and without color

Perform Treadmill Stress Test according to Bruce Protocol

Obtain Post Test Images
- Parasternal long axis, with and without color
- M-mode of mitral valve
- A4C, with and without color
- A5C, with and without color
- A3C, with and without color

Symptoms or arrhythmias?
- Yes
  - Document and end test
  - Continue test

- No
  - Continue test

Symptoms of BP drop?
- Yes
  - Document and end test

- No
  - Continue test

LVOT Gradient
Continuous Wave Doppler through the LVOT/aortic valve
- Apical 5 and apical 3 chamber views
- At rest and during Valsalva

Mitral Regurgitation Gradient
Continuous Wave Doppler
- Apical 4 and apical 2 chamber views
- At rest and during Valsalva

Tachycardia?
- Yes
  - Document and end test

- No
  - Continue post test imaging

LVOT Gradient
Continuous Wave Doppler through the LVOT/aortic valve
- Apical 5 and apical 3 chamber views
- At rest and during Valsalva

Mitral Regurgitation Gradient
Continuous Wave Doppler
- Apical 4 and apical 2 chamber views
- At rest and during Valsalva

Allow HR to slow to obtain clearer images and gradients

Yes
- No
- Continue test

Document and end test
1. Initial Localizing sequence/3 plane scout
   - Sagittal Plane
   - Coronal Plane
   - Axial Plane
2. Vertical long axis (VLA)/2 chamber scout
3. Horizontal long axis (HLA)/4 chamber scout
4. Short axis (SAX) scout
5. Short axis (SAX) cine stack:
   - Whole # slice shift position
   - 6mm/4mm Gap
6. Long axis (LAX) cines (use short axis cine stack to prescribe long axis views)
   - 3 chamber
   - 4 chamber
   - 2 chamber
   - Left ventricular outflow tract (LVOT) – 3 chamber (LA-MV-AOT)
   - RV inflow view
   - RVOT views
7. Hi resolution aortic valve
   - Small FOV
8. Hi resolution mitral valve imaging
   - A1P1, A2P2, and A3P3
9. Tagged cines of mid short axis and 4 chamber
10. Pre-contrast T1 and T2 MOLLIs
    - Mid short slice above mid SAX and 4 chamber
    - Dependent on the relaxation of protons in the magnetic field
11. HASTE (Half-Fourier Acquisition Single-shot Turbo Spin Echo) &
    - TRUFI (TRUe Fast Imaging with steady state precession) free breathing chest survey
    - 40-50 slices; 5mm/0mm Gap
12. Stress and rest perfusion on all patients with HCM
13. Gadolinium administration
    - 0.15 mmol/kg total dose given at 1 mL/sec followed by 20 mL normal saline flush
    - Contraindicated with renal failure or allergy to contrast agent
    - Record volume and time of injection
14. Perform phase contrast imaging
    - Aorta
    - Pulmonary Artery
    - Mitral Valve
    - Plane phase contrast imaging of the 3 chamber view to demonstrate flow acceleration in LVOT in HCM patients
15. Late Gadolinium Enhancement sequences
    - Single shots (short and long axis views)
    - Segmented acquisition (short and long axis views)
16. TI 600 msec imaging of LV short axis and 2 chamber stack with 0 Gap
17. Post contrast T1 MOLLIs (copy the pre-contrast images
Hypertrophic Cardiomyopathy Protocols

Exercise
Consider EP consultation to help assess risk of SCD, and to engage in shared decision making regarding ICD implantation for primary or secondary prevention of SCD.

Do NOT implant ICD to allow participation in exercise or athletics (Class III recommendation).

Assessment of risk for SCD with exercise in patients with HCM

- The overall risk of SCD with exercise in a given patient with HCM is low, but it is still roughly ten-fold higher than the risk in the general population. Current guidelines do not recommend use of algorithm or specific risk factors to direct recommendations for participation or exclusion from exercise or sport but recommend a complete assessment of risk, and shared decision making between expert clinician(s) and stakeholders including the patient and family, as well as third party interests.

Assessment might include H&P, TTE, ETT/Echo, Cardiac monitor, and CV-MRI

Some findings associated with increased risk of SCD with exercise in HCM (bold findings are higher risk)
- Unexplained syncope
- Family h/o SCD
- NSVT
- Severe LVH (>30 mm)
- LV dysfunction (LVEF <50%)
- Apical aneurysm
- LGE > 15% (especially if non-septal)
- Age<35 years
- Male gender
- Symptoms (pre-syncpe, DOE, CP)
- LAE
- Atrial Fibrillation
- Hypertensive response to exercise
- Exercise-induced dysrhythmia
- Resting LVOT gradient >30 mmHg
- Exercise LVOT gradient >50 mmHg

Consider the numerous benefits of exercise
- Improved prognosis
- Improved QOL
- Improved emotional well-being
- Lower risk of cancers
- Improved metabolic profiles
- Improved cardiorespiratory fitness
- Improved physical functioning
- Reduced overall CV risk
- Weight control
- May improve LV remodeling
- May improve the natural course of HCM

Some patient – specific considerations
- Patient’s goal(s)
- Patient’s risk tolerance
- Risk of anticoagulation
- Co-morbidities
- Occupational risk

Some sport – specific considerations
- Consequence of syncope (swimming, mtn. climbing, etc.)
- Adverse environmental conditions (extreme temps)
- Duration of exercise
- Avoidance of “dehydration”
- Location of exercise (isolation?)
- AED and/or EAP in venue?
- Aggressive “start-stop” sports?

High-intensity recreational, or moderate-to-high intensity competitive sports*
- Consider consultation with Sports Cardiologist
- Shared decision making about exercise
- Reassess annually, and for new symptoms

Low or moderate intensity exercise

*High intensity exercise can refer to >6 METS, HR > 70% MPHR, or perceived exertion >15 (Borg Scale)
Hypertrophic Cardiomyopathy Protocols

Cardiac Myosin Inhibitors
### Inclusion Criteria
- Symptomatic Obstructive HCM
- Resting or Exercise Gradient > 30 mmHg ON maximally tolerated GDMT (BB or Ca Blocker)
- NYHA Class 2 – 3 symptoms
- Female patients must not be pregnant or lactating and, if sexually active, must be using highly effective birth control method
- LVEF by Echo within 3 months of > 55%

**Refer to HCM Specialist for further screening and enrollment into REMS program**

### HCM Specialists
- Kenneth Taylor, MD
- Andrew Darlington, DO
- Rahul Loungani, MD
- Rajeev Singh, MD
- Charles Campbell, MD
- Matthew Crim, MD
- Denise Reedus, NP
- Julie Kim, NP
- Anna Lynch, RN

### Exclusion Criteria
- Known infiltrative or storage disorder causing cardiac hypertrophy that mimics HCM, such as Fabry disease, amyloidosis, or Noonan syndrome with LV hypertrophy
- Current or planned treatment with Verapamil or Diltiazem will need to be discussed with HCM specialist and dose reduction may be needed.

Follow up with General Cardiologist