

## Top Ten Things To Know HCM Guidelines

1. HCM is the most common genetic cardiovascular disease.
2. Over 600,000 people are affected by HCM in the U.S.
3. The purpose of these Guidelines is to review and provide consensus on the most important diagnostic and genetic tools used to treat HCM.
4. The nomenclature describing patients as “Noonan syndrome with left ventricular (LV) hypertrophy” or “Noonan syndrome with cardiomyopathy” is recommended in those patients with
  - (1) overt disease expression (with LV hypertrophy) appears to be confined to the heart and
  - (2) the definitive mutation is either one of a gene encoding proteins of the cardiac sarcomere or alternatively when the genotype is unresolved using current genetic testing.
5. The following are recommended diagnostic tools:
  - Genetic testing and counseling.
  - 12 lead ECG
  - Transthoracic echocardiogram
6. HCM patients and their first degree relatives should undergo genetic testing and counseling.
7. The 12 lead ECG is recommended to evaluate patients with HCM and 1st degree relatives, in addition every 12-18 months for adolescent 1st degree relatives with HCM who have no evidence of hypertrophy.
8. A transthoracic echocardiogram (TTE) is recommended
  - as an initial evaluation of patients who may have HCM.
  - to assess the effects of invasive therapy for obstructive HCM.
  - every 1-2 years in HCM patients to evaluate myocardial hypertrophy, dynamic obstruction and myocardial function.
9. The following are recommended treatments for HCM:
  - beta blockers for the symptoms of obstructive and nonobstructive HCM
  - verapamil for patients not responding to beta blockers
  - intravenous phenylephrine (or other pure vasoconstricting agents) for patients with acute hypotension in patients with obstructive HCM not responding to fluid
  - a permanent pacemaker should not be implanted in asymptomatic HCM and symptom controlled patients.
10. Although much progress has been made in understanding HCM more research is needed to present more opportunities to improve patient outcomes.