The New Hypertrophic Cardiomyopathy Practice Guidelines

Written by Maria Vinall

New practice guidelines for hypertrophic cardiomyopathy (HCM) were issued in 2011 by the American College of Cardiology Foundation/American Heart Association (ACCF/AHA) [Gersh BJ et al. *Circulation* 2011].

Michael J Ackerman, MD, Mayo Clinic, Rochester, Minnesota, USA, discussed the new guidelines for genetic testing for HCM from both the ACCF/AHA and from the Heart Rhythm Society/European Heart Rhythm Association (HRS/EHRA) [Ackerman MJ et al. *Heart Rhythm* 2011.

Under the ACCF/AHA guidelines genetic testing:

- is recommended in patients with atypical clinical presentation of HCM or when another genetic condition is suspected (Class I; Level of Evidence [LoE] B)
- is reasonable in the index patient to facilitate identification of first-degree family members who are at risk for developing HCM (Class IIa; LoE B)

Using the HRS/EHRA guidelines (both Class I; LoE C [ie Expert Consensus]):

- comprehensive or targeted HCM genetic testing is recommended for any patient in whom a cardiologist has established a clinical diagnosis of HCM based on examination of the patient's clinical history, family history, and electro-/echocardiographic phenotype
- mutation-specific genetic testing is recommended for family members and appropriate relatives following the identification of the HCM-causative mutation in an index case

Milind Y. Desai, MD, Cleveland Clinic, Cleveland, Ohio, USA, presented a summary of the recommendations for the use of echocardiography. Some of the specific recommendations concerned the use of transthoracic echo as part of:

- the initial evaluation of all patients with suspected HCM (Class I; LoE B)
- the screening algorithm for family members (Class I; LoE B)
- periodic screening for children of HCM patients (Class I; LoE C)

Martin S. Maron, MD, Tufts Medical Center, Boston, Massachusetts, USA, presented the recommendations for the use of cardiac MRI (CMR). The new guidelines indicate the use of CMR imaging:

- in patients with suspected HCM to detect segmental areas of increased left ventricular wall thickening that are not seen by echocardiography (eg, anterolateral wall, apex and posterior septum; Class I; LoE B)
- in patients with HCM when this additional information might have an impact on risk assessment and treatment decisions regarding appropriate selection of invasive septal reduction therapy (Class I; LoE B)
- for identification of high-risk patients using late gadolinium enhancement in selected HCM patients in whom risk stratification remains uncertain after assessment with conventional sudden death markers (Class IIb; LoE C)

Perry M. Elliott, MD, The Heart Hospital, University College London, London, United Kingdom, presented some of the new recommendations concerning the symptoms in HCM, such as chest pain, dyspnea and arrhythmia that may be explained by decreased myocardial perfusion and the resultant myocardial ischemia:

- assessment of coronary anatomy with computed tomographic angiography is reasonable for HCM patients with chest discomfort and a low likelihood of coronary artery disease (CAD) to assess for possible concomitant CAD (Class IIa; LoE C)
- the assessment of ischemia or perfusion abnormalities suggestive of CAD with single-photon emission computed tomography (SPECT) or positron emission tomography (PET) myocardial perfusion imaging (MPI) is reasonable in patients with HCM with chest discomfort and a low likelihood of CAD to rule out possible concomitant CAD (Class IIa; LoE C)
- recommendations do not support the use of routine SPECT MPI or stress echocardiography for detection of "silent" CAD-related ischemia in patients with HCM who are asymptomatic; nor is the use of PET to detect the presence of microvascular ischemia recommended for the prognosis of HCM (Class III; LoE C)

Overall there is a low annual rate of mortality from sudden cardiac arrest due to HCM. However, unacceptable death rates in the HCM population remain even in those being treated with medications, explained Matthew W. Martinez, MD, Lehigh Valley Health Network, Allentown, Pennsylvania, USA. Implantable cardioverter-defibrillators (ICDs) are a useful treatment option in patients who are at high risk of SCD, defined as young patients with a positive family history of SCD, unexplained syncope, left ventricularwall thickness >30 mm, associated CAD, nonsustained ventricular tachycardia, exercised-induced hypotension, or fibrosis detected by MRI (Class Ib; LoE B).

Paul Sorajja, MD, Mayo Clinic, Rochester, Minnesota, USA, comparing the outcomes from myectomy and septal ablation, reported that myectomy is the recommended standard treatment for HCM (Class IIa; LoE C), as it offers low immediate post-operative risk, a >95% chance of symptom relief and superb long-term survival benefits. Septal ablation works well if patients are carefully selected (Class IIa or IIb; LoE B) but has higher acute complications, often including heart block, hence the need for a pacemaker following ablation. The two procedures appear to be comparable in terms of gradient relief, symptom relief, and early survival; however, there continues to be concern regarding the potential long-term consequences of the ablation-induced infarction, including data from ICD monitoring that suggest an increased risk of ventricular arrhythmia among HCM patients post ablation compared with septal myectomy. A definitive randomized trial that compared long-term outcomes between these two procedures has yet to be performed.

New Anti-Diabetes Agents Offer Promise in the Fight Against CVD

Written by Rita Buckley

Type 2 diabetes mellitus (T2DM) is a major risk factor for ischemic heart disease, and cardiovascular disease (CVD) is the leading cause of morbidity and mortality for individuals with T2DM [McEwen LN et al. *Diabetes Care* 2012]. CVD is also the largest contributor to direct and indirect medical costs that are associated with T2DM. Common conditions that coexist with T2DM (eg, hypertension and dyslipidemia) are clear risk factors for CVD; however, a diagnosis of T2DM itself confers independent risk [Whittington HJ et al. *Cardiol Res Pract* 2012].

Numerous studies have demonstrated the efficacy of targeting and controlling individual CV risk factors (eg, blood pressure less than 130/80 mm Hg, low-density lipoprotein cholesterol less than 100 mg/dL, HbA1C <7%) in preventing or slowing the progression of microvascular and macrovascular disease in patients with T2DM [American Diabetes Association Standards of Medical Care in Diabetes—2012. *Diabetes Care* 2012] (Figure 1). Larger benefits are seen when multiple risk factors are globally addressed in patients with T2DM [Buse JB et al. *Diabetes Care* 2007; Gaede P et al. *N Engl J Med* 2008].

However, randomized clinical trials have also suggested the limits of intensive CV risk factor control in T2DM [The ACCORD Study Group. *N Engl J Med* 2010; Duckworth W et al. *N Engl J Med* 2009; ADVANCE Collaborative Group. *N Engl J Med* 2008]. In particular, achieving intensive glucose control alone may be insufficient to reduce major CVD events. A new medication class that may reduce CVD in patients with T2DM uses molecules that activate the incretin system to raise or mimic glucagon-like peptide-1 (GLP-1). In a recent review, Motta et al. [*Recent Pat Cardiovasc Drug Discov* 2012] reported that incretin-based agents improve glycemic control by mechanisms