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### Hypertrophic cardiomyopathy

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In recent years a major leap forward has been made in diagnostic and therapeutic options available for patients with HCM. In chapter 2, recently developed diagnostic options and their current and potential use in patients with HCM are reviewed. A large number of mutations in the  $\beta$ -myosin heavy chain gene and some other genes have been identified that cause HCM. Although it is today almost impossible to diagnose HCM using routine genotyping in an individual patient, the genetic abnormality can be detected in about half of the families with HCM using linkage studies. The results of genetic screening in such families have shed new light on the value of both the standard electrocardiogram and echocardiography in screening for HCM. A significant overlap of electrocardiographic and echocardiographic findings is present between genetically affected individuals and normal controls. Analysis of heart rate variability and assessment of late potentials appear not useful in diagnosing HCM or detecting high risk patients. The role of echocardiography in the management of patients with HCM has increased tremendously. Several structural abnormalities of the mitral valve leading to different therapeutic strategies can be detected. Intraoperative transesophageal echocardiography can be very helpful in decision making and in guiding the intervention. The role of QT interval dispersion, magnetic resonance imaging and positron emission tomography are also reviewed in chapter 2, but these subjects are discussed in greater detail in chapters 4 and 6-8.

In chapter 3, novel therapeutic options are discussed. The preferred medical therapy for HCM is still verapamil, although  $\beta$ -blockers and disopyramide also have a role in the treatment of patients with HCM. In recent years, to non-pharmacologic treatment modalities other than surgery have been developed that both have in common an induction of a remodelling process of the left ventricle. Outflow tract obstruction can be eliminated by injecting absolute alcohol into the first major septal artery, but major concern exists on the long term effects of this method. Permanent dual chamber pacing improves subjective symptoms in about 80 % of patients with drug refractory obstructive HCM, but again it is not clear if this therapeutic option might show detrimental on the long run.

In chapter 4, the diagnostic value of spin-echo magnetic resonance imaging  $\nu$  transthoracic echocardiography is compared. In 52 patients with HCM 2 hypertrophy scores are assessed with both techniques. The hypertrophy scores correlate well between echocardiography and magnetic resonance imaging, but echocardiography is of insufficient quality for calculating adequate hypertrophy scores in a substantial part of the patients. magnetic resonance imaging provides the most comprehensive diagnostic information in patients with HCM.

In chapter 5, the effects of acute intravenous  $\nu$  long-term oral verapamil administration on diastolic function are noted. Both modes of administration have distinctly different results with respect to diastolic function. After oral verapamil administration, left ventricular diastolic function improved in most patients, but after intravenous administration no consistent improvement was noted. It is not possible to pre-

dict the effect of long-term oral verapamil with an intravenous test dose in an individual patient with HCM.

In chapters 6 and 7, the effects of permanent dual-chamber pacing on myocardial perfusion in patients with HCM are noted. Pacing results in clinical improvement and a reduction of anginal complaints. During pacing, a decrease of resting left ventricular myocardial blood flow and blood flow during pharmacologically induced coronary vasodilation is found. Global perfusion reserve is unchanged, but myocardial perfusion reserve shifts into a more homogeneous distribution. These effects of pacing are probably caused by a reduction in anteroseptal fiber strain and oxygen demand due to early septal activation.

In chapter 8, QT dispersion and the occurrence of late potentials are assessed. The incidence of late potentials in patients with HCM is low. QT dispersion is increased and QT interval prolonged but both are not related to the occurrence of ventricular arrhythmias. The electrocardiographic manifestations of delayed conduction and abnormal repolarisation are probably blunted in patients with septal forms of HCM due to the early activation of the septum.

In conclusion, the most promising diagnostic tools in patients with HCM are genetics, magnetic resonance imaging and positron emission tomography. These techniques may add additional information necessary for a reliable risk stratification in patients with HCM. Of the therapeutic options the effects of dual-chamber pacing are hopeful, but cautious optimism is warranted until more light is shed on the exact effects of pacing on the pathophysiology and on the long-term effects.