Hypertrophic Cardiomyopathy (HCM)

Hypertrophic cardiomyopathy (HCM) is an inherited condition that leads to abnormal thickening of the heart muscle, most often of the left ventricle (the main pumping chamber of the heart). The thickened muscle reduces the efficiency with which the heart works. People with HCM may experience chest pain, shortness of breath, dizziness, fainting episodes or palpitations, but many people with HCM never have any symptoms. Tragically, in some cases the first sign of the condition is sudden death, especially in young athletes. Therefore, it is very important that everyone with a family history of HCM is screened by a cardiologist.

About 1 in 500 people have HCM.

In adults, the walls of the left ventricle are usually 7mm to 10 mm thick (can be thicker at post mortem/autopsy). In HCM, the walls are more than 13 mm thick and the thickness can vary in different parts of the ventricle. The wall thickening is often unevenly distributed, unlike in people with high blood pressure. This is illustrated in the diagram below. In about 25% (1 in 4) of people with HCM the thickened muscle obstructs the flow of blood out of the heart – this is known as the obstructive type of hypertrophic cardiomyopathy (sometimes referred to as HOCM).

How it is inherited
HCM is usually inherited as an autosomal dominant condition. See section on Genetic Inheritance for more information.

Genetic testing
Genetic testing is now commercially available and involves screening 10 genes that can cause the condition. The gene alteration will be identified in approximately 50-60% of families.

Treatment
Most people with HCM will not require any specific treatment but should avoid competitive high-impact sports. There are medications that can help those with symptoms. A small
proportion of patients, who are at highest risk of an abnormal heart rhythm as assessed by a cardiologist, will benefit from a pacemaker-defibrillator (known as an ICD or implantable cardioverter defibrillator). This is a device that will detect an abnormal rhythm and deliver a ‘shock’ to correct the rhythm (see section on ICD for more information).

People with an obstruction to blood flow in the heart (HOCM) may need to have the obstruction reduced through surgery (myectomy or alcohol septal ablation). These procedures are not necessary for everyone and whether they are appropriate for you should be discussed with your cardiologist.

Your risk of developing complications may increase with age so long-term cardiology follow-up is essential. Everyone with HCM should see their cardiologist at least once a year.

For more information

Further Information is available from the Cardiomyopathy Association of Australia at www.cmaa.org.au

You can also find out more about genetic heart conditions and the latest research news by visiting the Australian Genetic Heart Disease Registry at www.heartregistry.org.au or the Cardiac Inherited Diseases Group (New Zealand) at www.cidg.org