

# Appendix A: Cardiovascular conditions and their treatments

## Congenital heart disease

Congenital heart disease refers to any malformation or disease of the heart present from birth. It includes structural defects, congenital arrhythmias and some cardiomyopathies. Over 90% of cardiac pathology in the paediatric population in the developed world is congenital in origin, in that it is present at birth, even if undetected until older; in contrast, adult heart disease is largely classified as being acquired. Acquired heart disease develops after birth and examples of heart disease developed in childhood include most cardiomyopathies and inflammatory heart disease such as rheumatic heart disease and Kawasaki Disease.

The diagnosis and treatment of heart malformations has dramatically improved over the past few decades, with major advances in both surgical and percutaneous transvascular techniques (transcatheter 'keyhole techniques'). Examples of the latter include balloon dilation for valve stenosis and device closure of the arterial duct, secundum atrial septal defect and certain ventricular septal defects.

Surgical risks are highest for neonates who present in poor condition so a goal of congenital heart disease services is to spot heart disease as early as possible, ideally before birth (referred to as antenatal diagnosis).

Services for congenital heart disease are concentrated in a small number of centres to ensure that there is a sufficient number of procedures undertaken to develop and retain skills, experience, and organisational processes. There is also a need for them to be in close proximity to other specialist tertiary services, including the care of children with acquired heart disease.

## Acute coronary syndromes (including heart attacks)

The most common form of heart disease in adults relates to coronary artery disease (atherosclerosis or 'hardening of the arteries'). This may result in angina or acute coronary syndromes such as unstable angina or heart attacks when the build-up of fatty deposits within the wall of the artery leads to blockages that affect the flow of blood to the heart muscle. Sometimes, inflammation around a cholesterol-rich fatty deposit causes an ulcer or fissure of the inner surface of the artery; the body's automatic healing response is associated with the development of blood clots – clots that serve to heal the fissure but which may also cause an abrupt cessation of blood flow, leading to heart attacks. Less commonly, heart attacks can occur through other mechanisms including spontaneous coronary artery dissection or due to adrenaline surges ('Takotsubo syndrome'). Whatever the mechanism, a heart attack can be associated with sudden changes in heart rhythm,

some of which can be lethal – some, but not all, heart attacks can lead to cardiac arrest. For survivors, the heart muscle may be considerably weaker than before and patients may be prone to heart failure (see below).

Significant improvements in treatment have occurred over the last few decades, with drugs aimed at stabilising atherosclerosis, reducing the risk of heart attacks. The risk of heart attacks can also fall dramatically with better lifestyles (avoiding smoking, taking exercise and having a healthier diet).

When a major heart attack occurs, it is essential to restore blood flow down the coronary artery and the best way to do this is with emergency angioplasty (so-called 'primary PCI', or PPCI, or just 'primary angioplasty'). Once a patient calls for help, speed is of the essence as the longer it takes to restore blood flow the more damage is done to the heart and the more likely the patient is to have a lethal rhythm disturbance and later heart failure.

Following a heart attack, drug treatments to thin the blood (anti-platelet drugs) and so reduce the likelihood that further clots will obstruct the coronary artery, as well as ones which reduce cholesterol levels (statins and others), control blood pressure, stabilise heart rhythm, maintain heart pump function and reduce inflammation (beta blockers, angiotensin-converting enzyme inhibitors (ACE-Is) and others) have all led to improved outcomes. Patients' well-being and longer term outcomes are also improved by cardiac rehabilitation programmes.

In this report, the terms 'higher-risk' and 'lower-risk' have been used to differentiate those patients whose heart attacks are characterised by a specific electrical change seen on an electrocardiogram (ECG) early after the onset of symptoms from those in whom it is not. Those with ST-segment elevation are most likely to have complete coronary occlusion and require primary angioplasty. Higher-risk heart attacks in this report therefore refer to what clinicians call ST-elevation myocardial infarction (STEMI) – patients who are at high risk of substantial heart muscle damage or early death. Patients who do not have this ECG change, who are likely to have only partially obstructing clots in the coronary artery and so do not need immediate angioplasty, are at lower risk of early death. Lower-risk heart attacks in this report therefore refer to what clinicians call non-ST-elevation myocardial infarction (NSTEMI) – though, because patients with NSTEMI tend to be significantly older and have more comorbid conditions than those with STEMI, over a one-year period the risk of death is about the same.

## Percutaneous coronary intervention (PCI) or 'angioplasty'

When obstructions in the heart arteries lead to exertion-

induced chest pain (angina) that cannot be controlled by medical treatment, then patients may be helped by methods to improve blood flow. The two techniques are percutaneous coronary intervention (PCI) (often referred to as 'angioplasty') and coronary artery bypass grafting (CABG). With angioplasty, a fatty deposit is pushed aside by the use of a balloon or wire mesh ('stent') that can be inserted under X-ray vision. The stent is taken up to the heart through a guide catheter that is passed into the body under local anaesthetic from either the groin (using the femoral artery) or the wrist (using the radial artery). Recent research has shown that complications are fewer when the wrist is used.

When angioplasty was first introduced, the arterial narrowing was stretched with just a balloon, but sometimes problems led to the need for emergency open heart surgery. Even successful treatments did not hold up over time because the vessel could re-narrow for a number of reasons. These early complications and the subsequent potential for re-narrowing were significantly reduced by the use of stents. The first stents were tubular wire meshes ('bare metal stents') but even some of these re-narrowed because of scar tissue developing inside the stent. Research led to the development of stents with a plastic coating which contained special drugs to minimise the development of scar tissue ('drug-eluting stents'). The drug is slowly released into the wall of the vessel to have its effect. Although there was a slight concern that drug-eluting stents might be a little more prone to the developments of clots after implantation ('stent thrombosis'), improvements in technology and the routine use of dual anti-platelet therapy (DAPT – using two drugs that inhibit platelet function by different mechanisms, thus enhancing the effect that can be achieved with just one drug) have led to improved outcomes. New research has led to the development of plastic coats on these stents that slowly dissolve over time (bioabsorbable polymers) which may further improve outcomes. Additional research is also being done to develop stents made entirely of special plastics that will slowly dissolve away (bioabsorbable stents).

In the early years of using angioplasty, it was used mainly for patients with stable angina. However, over the last 20 years it has been used more and more to treat patients with acute coronary syndromes, and especially for patients with heart attacks.

## Heart surgery

The most common form of heart surgery is coronary artery bypass grafting (CABG), which is used for patients with severe angina or after a heart attack. There is good evidence for the benefits of CABG, which may also improve heart pump function in some patients with heart failure secondary to blocked blood vessels. As the specialty of cardiology developed, more and more people were put forward for heart surgery and waiting times for treatments grew to unacceptable levels. However, from 2000 onwards, much work has been done to improve access to heart surgery and these waits have fallen dramatically, although there is continuing awareness of variation around the country. Although open heart surgery

is classified as major surgery, new 'minimally invasive' techniques have been introduced, recovery in general is quicker than it used to be, patients can be discharged earlier and complications have fallen. There are some major complications associated with all of these cardiac treatments but fortunately they are infrequent. Outcomes in terms of improved symptoms and quality of life are well established.

Open heart surgery is also required for patients with severe heart valve lesions causing the valve to be narrowed or very leaky. These problems can lead to patients becoming tired and breathless and may result in irreversible heart muscle weakness or changes in the lungs. These valve abnormalities may be due to congenital abnormalities, rheumatic heart disease (less frequent in the UK nowadays) and other inflammatory conditions affecting the valves, or to degenerative problems causing valves to thicken or split over time. It is important to make accurate diagnoses and to follow patients up so that treatment can be provided before these changes occur. Valve replacements make up a large part of the workload of surgical programmes although new techniques mean that many of these valve problems can now be treated by surgical repairs. For patients with narrowing of the aortic valve but who are at high risk for surgery, a new technique called transcatheter aortic valve implantation (TAVI) has been introduced, whereby a new valve can be inserted without the need for open heart surgery. Other new techniques for valve problems and other structural abnormalities of the heart are being explored.

Open heart surgery may also be needed for rare congenital problems that might not present until adulthood as well as for problems with the aorta, the major artery through which blood passes from the main heart pump to the rest of the body. This can weaken because of congenital problems with the strength of the vessel wall or through wear and tear associated with high blood pressure, atherosclerosis and ageing.

## Heart failure

Whether due to congenital heart muscle abnormalities (cardiomyopathies), inflammation of the heart (myocarditis) or damage associated with problems arising from coronary artery or valve disease, the pumping chambers of the heart may increase in size and their pump power reduce ('heart failure with reduced ejection fraction' or HFrEF). This may be associated with fatigue and breathlessness and may be associated with dangerous heart rhythm abnormalities and a reduced survival rate. The ejection fraction is a measure of the pumping capability of the heart. Symptoms might also occur in patients with thickened heart muscle that may become stiff. Although the pump power may be retained, the wall of the pump does not relax well, the cavity of the main heart chamber can reduce in size and this leads to back pressure on the blood vessels in the lungs. The syndrome of heart failure can be exactly the same but this combination is referred to as 'heart failure with preserved ejection fraction' or HFpEF.

In the past, symptoms of heart failure could only be improved by the use of diuretics ('water tablets') and in some people

by the use of digoxin, but over the last two decades new treatments have had an impact on reducing the rate of deterioration of heart muscle problems, have made patients less prone to dangerous heart rhythm abnormalities and have helped improve symptoms and quality of life. These 'disease-modifying treatments' include beta blockers, ACE-Is, angiotensin receptor blockers (ARBs) and mineralocorticoid receptor antagonists (MRAs).

In some patients with specific characteristics related to a combination of their symptoms, their pump power and the shapes seen on their ECGs, the power of the pump may be improved by special pacemaker devices (cardiac resynchronisation therapy or CRT). These devices may also be able to monitor the patient's heart rhythm and provide special pacing techniques or shock treatment should any life-threatening rhythms occur. Other devices provide these functions but without the resynchronisation function – so-called implantable cardioverter defibrillators (ICDs). Most of the research for improved outcomes has been on patients with HFrEF. Ongoing research is looking to see whether new treatments other than that aimed at the causative mechanism might improve outcomes for patients with HFpEF.

## Cardiac rhythm management

Patients may suffer from a number of problems should they develop abnormalities of heart rhythm, whether a conduction problem leading to the heart beating too slowly or electric circuit problems that can cause the heart to pump too fast. Although a small number of patients present with slow heart rates due to congenital abnormalities of the conducting system of the heart, most of these problems occur in older adults as the electric conduction pathways become scarred with age. Patients with heart block are prone to fatigue, breathlessness, black-outs and even sudden death. Their symptoms and prognosis can be dramatically improved by the implantation of a pacemaker. Research suggests that better outcomes in terms of symptoms occur when the electrical coordination between the

upper chambers of the heart (the atria) and the lower pumping chambers of the heart (the ventricles) is maintained (so-called 'physiological pacing'). This usually requires a pacemaker with one lead in the right upper chamber and one lead in the right lower chamber of the heart (dual-chamber pacing).

Some arrhythmias are not especially dangerous but can cause impairment to quality of life by producing symptoms such as palpitations, dizziness, breathlessness and fatigue. There are subsets of patients though where these problems can lead to a weaker heart muscle or where the rhythm disturbance can degenerate into even faster and more dangerous rhythms. Many of the problems seen in children and young adults may arise as a consequence of residual pathways ('short-circuits') that may be brought into play in certain circumstances. There are some rare congenital causes of dangerous heart rhythm problems which lead to the pumping chambers of the heart going dangerously fast (ventricular tachycardia or ventricular fibrillation), although the vast majority of patients with these two sorts of rhythm abnormality have scarred hearts associated with problems such as a previous heart attack or cardiomyopathy.

It is important for patients with these rhythm problems to be seen by a cardiologist with expertise in their management (electrophysiologists). The correct diagnosis is not always possible from looking at a standard ECG and special internal recordings of the heart rhythm might be needed (an electrophysiology or EP study). Once the correct diagnosis is made, the heart rhythm team can decide whether this is best treated with special anti-arrhythmic drugs, ablation techniques or pacemakers or the special implantable devices mentioned above under 'Heart failure', namely ICDs and CRT devices. The latter devices have been shown to be better at improving survival rates than using anti-arrhythmic drugs, although for some patients the use of drugs is all that is needed. Ablation techniques are designed to interrupt either the initiating areas or the circuits that can maintain these rhythms, and in many patients can result in a cure for the rhythm disturbance.