Percutaneous Intervention in Adult Patients with Congenital Heart Disease - An Overview

Dr. Boron CW Cheng

MBBS (HK), MRCP (UK), FHKAM (Medicine)
Associate Consultant, Division of Cardiology, Department of Medicine, Queen Elizabeth Hospital

The field of interventional cardiology for adult congenital heart problems is evolving rapidly. Both native and postoperative lesions can be tackled with catheter interventional techniques. This overview will cover the most commonly encountered clinical problems.

The most common interventional procedures for closure of intracardiac communications in adults are the closure of atrial septal defect (ASD) and patent foramen ovale (PFO). Other defects include muscular ventricular septal defect (mVSD), and perimembranous ventricular septal defect (pmVSD). Closure of postoperative intracardiac communications is included in this category. (e.g. interatrial communication in post-Fontan patients)

Other communications, which can be tackled with catheter interventional techniques includes patent ductus arteriosus (PDA) and coronary artery fistulae (CAF). Vessels or conduit narrowing, for example, coarctation of aorta (COA) and pulmonary artery stenosis, can also be tackled with interventional techniques.

Atrial Septal Defect (ASD)

Early catheter closure in asymptomatic atrial septal defects with echocardiogram evidence of right ventricular enlargement is to reduce the risk of atrial arrhythmias, development of right ventricular failure symptoms and pulmonary vascular disease. If enlargement of the right ventricle is present, closure is indicated regardless of the presence or absence of symptoms. In the presence of raised pulmonary arterial pressure and left ventricular dysfunction, balloon test occlusion is advisable before definitive closure.

Percutaneous catheter closure of ASD is feasible in secundum type defects less than 40mm. Device closure is not feasible in sinus venous defects, atroventricular septal defects and secundum ASDs greater than 40mm.

The most widely used device nowadays is the Amplatzer septal occluder (AGA Medical, Golden Valley, Minnesota, USA). This is a double disk, self-centering design constructed from a meshwork of nitinol wire with a loose polyester fill. One disk is positioned in each atrium and they are joined by a waist that is sized to the defect stretched diameter and positioned across the defect. The size of the device is determined by the diameter of the waist, available from 4 to 40mm in diameter.

Atrial Septal Defect (ASD)

Fig. 1 Common types of Atrial Septal Defect

Fig. 2. An Amplatzer Septal Occluder

Fig. 3. Diagram showing the preferential of blood from IVC towards the fossa ovale. (Meier et al. Circulation 2003)
Cryptogenic stroke
A patent foramen is present in 27% of the population and has been implicated in young patients with cryptogenic stroke by allowing paradoxical embolisation of the clot that has developed either in the systemic veins or in the foramen tunnel itself. Under normal situations, the left atrial pressure exceeds that of the right atrium, thereby compressing the flap of the septum primum across the oval foramen in the left atrium. For a venous thrombosis to cross a patent foramen ovale, the pressure in the right atrium must be greater than that in the left atrium. Such changes occur transiently during Valsalva Manouvrres in daily life. It is therefore anticipated that closing a patent oval foramen will reduce the risk of recurrent embolic strokes. The diagnosis is usually made as the result of investigation of a stroke or transient ischaemic attack using echocardiography or transcranial Doppler.

Platypnea Orthodeoxia syndrome
This uncommon syndrome occurs when intracardiac shunting is present in the upright position leading to oxygen desaturation and breathlessness. This may occur in post-right ventricular infarct, thoracic aortic aneurysm or decrease in right atrial compliance secondary to aging. In this situation desaturated blood flows from the inferior vena cava across the foramen ovale into the left atrium, most commonly when the patient assumes a standing position. This uncommon condition can be resolved effectively by device closure.

There are many devices available for closure of PFO. The most common examples are the CardioSEAL septal occluder and the Amplatzer PFO occluder. Other newer devices include the Premere device.

Ventricular septal defect (VSD)
Device closure may be considered for highly selected cases of perimembranous and muscular VSD. Closure of VSD is indicated in patients with large intracardiac shunting, history of bacterial endocarditis and development of aortic regurgitation. A perimembranous VSD may be regarded as suitable for device closure if the distance from the superior edge of the defect to the insertion of the right coronary aortic valve leaflet is adequate. For Amplatzer perimembranous VSD device, the distance required is no less than 3mm. A muscular VSD may be closed with both the Amplatzer family device or the CardioSEAL occluder. Indications for closure of muscular VSD are similar to perimembranous VSD except for the development of aortic regurgitation.

Patent ductus arteriosus (PDA)
The rationale for closure of a PDA is the prevention of haemodynamic deterioration and elimination of the ductus as a substrate for bacterial endocarditis. Current recommendations are that any clinically apparent PDA should be closed.

At present, the most frequently used device for PDA closure in adults is the Amplatzer PDA occluder. It is particularly suitable for large diameter PDAs (>3mm).

For small ductus in adults or in patients with a small residual leak after a previous surgical or device closure, coils may be considered as an alternative option.

Coronary artery fistula (CAF)
Small coronary arteriovenous communications are not uncommonly observed during selective coronary arteriography in adults. These small communications are usually not significant, cause no haemodynamic sequelae, and hence may be ignored. In a few patients, there is a large communication between a coronary artery and a cardiac chamber, coronary sinus or pulmonary trunk. These coronary artery fistulae are characterised by enlargement of the coronary artery giving rise to the fistulous connection. The right coronary artery is the site of origin in more than 50% of cases, and the common sites into which the fistulae feeds are a cardiac vein, the right atrium and the right ventricle. Bilateral fistulae are present in up to 5% of cases. Run-off through a large fistula may lower intracoronary diastolic pressure and produce myocardial ischaemia by a steal phenomenon. Significant shunting from the left to the right may be present, resulting in chamber enlargement secondary to volume overload. Giant fistulae are at risk of rupture.

Multiple occlusion techniques including vascular plugs, coils and balloons, have resulted in complete occlusion in the majority of patients.

Coarctation of aorta (COA)
The mean survival of patients with untreated COA is 35 years, with a mortality rate of 75% by 50 years of age. Death in unrepaired patients is usually due to heart failure, coronary artery disease, aortic rupture or dissection, concomitant aortic valve disease, infective endocarditis or cerebral haemorrhage. COA is considered significant if the gradient is greater than 20mmHg between the arm and the leg with or without proximal systemic hypertension, or in the presence of upper extremity hypertension accompanied by echocardiographic or angiographic evidence of aortic obstruction.
stents may also reduce the incidence of aneurysm formation by limiting the amount of dilatation required and thus reducing the degree of traumatic injury to the aortic wall.

Pulmonary artery stenosis

The goals of intervention include a reduction of right ventricular pressure load, maintenance of right ventricular function, reduction in the amount of regurgitation through an abnormal pulmonary valve and increase in pulmonary perfusion in the affected lung. The most common site of pulmonary artery stenosis is found at the point of bifurcation of the main pulmonary artery. Branch pulmonary artery stenoses may reflect intrinsic abnormalities of the pulmonary arteries or may be the result of previous surgery such as systemic to pulmonary artery shunts.

Catheter intervention is again based on balloon and stent dilatation of the stenotic segment.

Conclusions:

The number of patients treated with interventional catheter techniques in the adult congenital cardiac clinic continues to grow. Many of these techniques have the benefit of avoiding surgical procedures involving sternotomy or thoracotomy. In experienced hands, most catheter interventional procedures have good safety profiles and high success rates.