Transcatheter Treatment of Congenital Heart Defects Using Amplatzer Devices

By Blaž Kosmač and Tomaž Podnar

Introduction

Congenital heart disease is the most frequent form of major birth defect, affecting nearly 1% of newborn babies. Surgery has been the only treatment option for a number of years. Since the 1970s, transcatheter devices have been available to occlude certain types of congenital heart defects, namely secundum-type Atrial Septal Defect (ASD II), Patent Foramen Ovale (PFO), Patent Ductus Arteriosus (PDA) and, most recently, Perimembranous Ventricular Septal Defect (PMVSD) and Muscular Ventricular Septal Defect (MuscVSD). These devices make possible an alternative to established surgical procedures. The occlusion of these intracardiac and extracardiac communications has been revolutionised by the development of Amplatzer occluders. These devices are made of heat-treated nitinol wire mesh formed into different shapes. At the University Medical Center in Ljubljana, Amplatzer devices have been used regularly since 2000, when the first closures of an ASD II were

Assistant Professor Tomaž Podnar, MD, Cardiology Unit, University Children's Hospital, University Medical Center, Vrazov trg 1, 1525 Ljubljana, Slovenia.

Blaž Kosmač , MD, Cardiology Unit, University Children's Hospital, University Medical Center, Vrazov trg 1, 1525 Ljubljana, Slovenia.

performed. A total of 171 procedures were performed by the end of 2005.

Closure protocol

All procedures are performed under general anaesthesia. Patients (both children and adults) are admitted to the hospital the day before the procedure. Provided there are no complications, they are discharged the day after the procedure. Physical examination, ECG, chest x-ray and echocardiogram are performed before the procedure and at follow-up evaluations. The procedures are performed through the groin. Under x-ray monitoring, catheters are inserted through the femoral vein and femoral artery into the heart and large vessels. Defects are precisely delineated by contrast injections and accurate measurements of the defects are performed. In addition, when closing ASD II, PFO or VSDs, an echo probe is placed in the oesophagus to monitor the procedure. An appropriately sized device is selected and positioned through long delivery sheaths introduced through the femoral vein into the defect. Patients are awakened immediately after the procedure. The follow-up protocol includes acetylsalicylic acid, up to 100 mg daily, for six months. Antibiotics during interventions are commonplace, and prevention against bacterial endocarditis is recommended for 6 months until the inserted device is completely covered by tissue.

Secundum-type atrial septal defect (ASD II)

The ostium secundum atrial septal defect is the most common type of atrial septal defect and comprises 6–10% of all congenital heart diseases. There is a low-pressure, left-to-right shunt across the ASD II which causes right heart dilation and pulmonary overcirculation. The indication for closure of the ASD II is clinical or echographic evidence of a significant left-to-right shunt causing dilatation of the right heart chambers. Patients suited to percutaneous closure have defects up to a maximum of 40mm in diameter with sufficient tissue rims (>5mm) surrounding the defect. Very large defects with incomplete tissue rims (except toward the aorta) require surgical closure.

The Amplatzer Septal Occluder (ASO) has the shape of two disks connected by a central stent-like connecting waist,
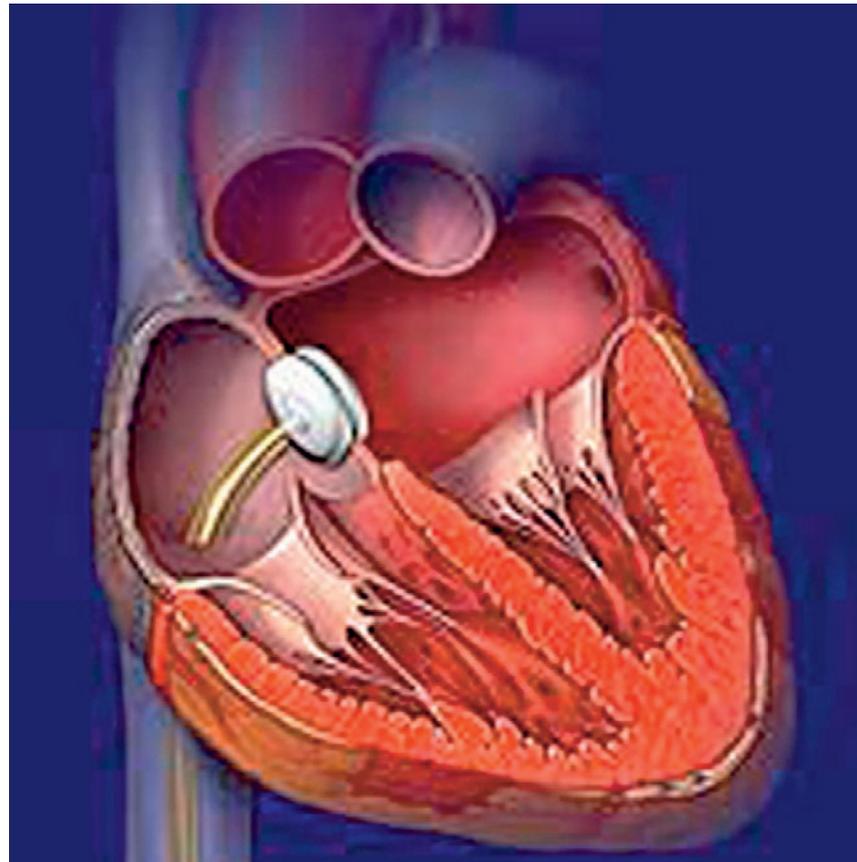
Figure 1: The three components of the Amplatzer Septal Occluder: left atrial disk, central waist and right atrial disk.

of various diameters to allow closure of both small and large defects (Fig. 1). The atrial septal defect is sized with a balloon catheter. An occluder of the correct size is selected and introduced into the left atrium via a long delivery sheath. The left atrial disk of the occluder is extended and pulled against the septum. The sheath is then pulled back to deploy the rest of the device – central waist and right atrial disk. After positioning, the device is assessed by transoesophageal echocardiography and then released (Figs. 2 and 3).

The defect is closed by inducing thrombosis on three polyester patches sewn into the device, which become covered by neointima within two months. Until the delivery cable is disconnected, the device can be withdrawn into the long delivery sheath and removed from the body. Immediate complete closure of the defect is observed in most patients, while in the vast majority of the others the residual atrial shunt disappears in the next few months. To date, no other device has achieved such high closure rates, which is attributable to the unique design features and increased thrombogenicity of the device.

Complications of percutaneous ASD II closure using ASO are exceptional and are less frequent than after surgical closure.

ASD occluder positioning in the heart.

Figure 2

Figure 3

Figures 2 and 3: Transoesophageal echocardiograms of an atrial septal defect measuring aproximately 20mm in diameter before (up) and after (down) occlusion with an Amplatzer Septal Occluder (LA=left atrium, RA=right atrium).
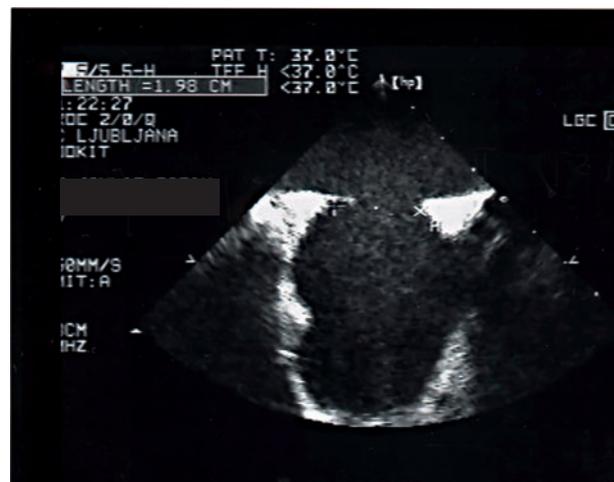
Patent Foramen Ovale (PFO)

The foramen ovale is essential in fetal circulation for blood flow across the foetal atrial septum. After birth, changes in right and left atrial pressures cause functional closure of the foramen ovale. The closure is complete by age two in about 75% of individuals. However, in the remaining 25% the fusion does not take place – the foramen remains patent (Fig. 4). The reasons why PFOs fail to close are unknown, but they likely relate to multifactorial inheritance. The defect is usually oblique and slit-shaped, resembling a tunnel. It is known that PFO carriers are at increased risk for several serious clinical syndromes, including paradoxic systemic embolism such as cryptogenic stroke, acute myocardial infarction, decompression sickness in divers and a rare platypnea orthodeoxia syndrome. It has also been associated with an increased prevalence of migraine, although the causal relationship between the two has yet to be determined. Studies on annual recurrences after a cerebral vascular accident or transient ischemic attack have reported an incidence ranging from 3–16%. PFO alone increases the risk of recurrent events 5-fold, with an even higher risk in the presence of an atrial septal aneurysm. According to various authors, transcatheter PFO closure with present techniques seems to protect against recurrent strokes in this patient population and is currently the only unequivocal indication for PFO closure.

Paradoxical embolisms are demonstrated by transcranial Doppler examination (TCD), while patency with consequent left-to-right and right-to-left shunting is shown by colour flow mapping in transoesophageal echocardiography.
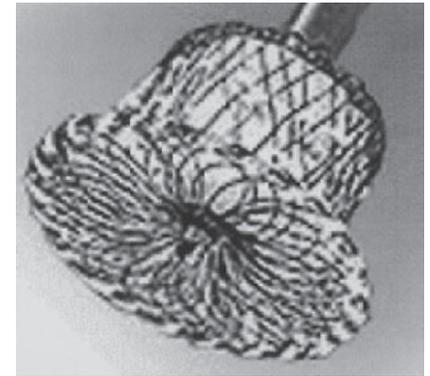
Figure 4: The Amplatzer PFO Occluder. The device differs from the atrial septal defect device in not having a central waist. Its right atrial disk is larger than the left atrial disk and is oriented in a concave direction towards the atrial septum.

Several PFO occluders are currently available on the market; however, Amplatzer PFO Occluders are most often used. The occluder has no central stent and is designed to close the flap-valve of the patent foramen ovale (Fig. 4). The procedure is similar to ASD II
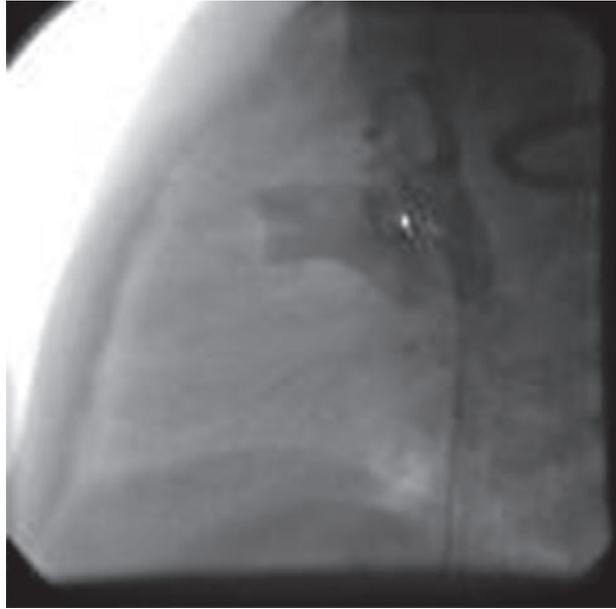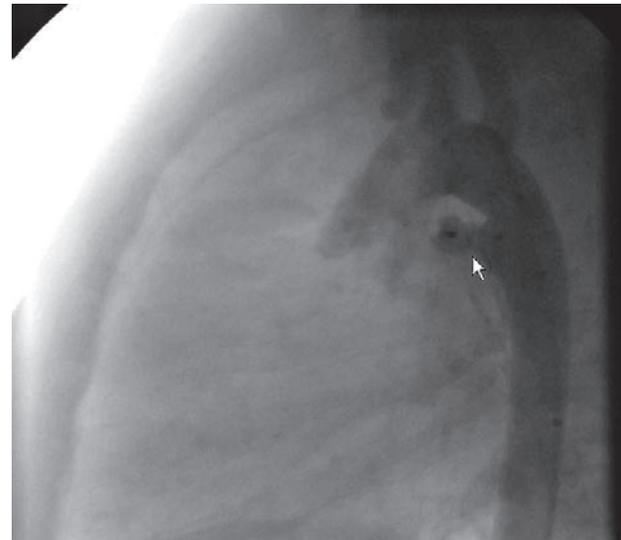

Figure 7: Aortogram showing a large PDA with a shunt of contrast from the aorta to the pulmonary artery.

Figure 8: The Amplatzer PDA Occluder is a self-expanding, mushroom-shaped device made from a 0.004-inch thick nitinol wire mesh. A thin retention disc, 4 mm larger in size than the diameter of the device, ensures secure positioning in the mouth of the PDA.

Figures 5 and 6: Colour flow mapping in transoesophageal echocardiography demonstrating spontaneous right-to-left shunting through the slit-like PFO (up). After device deployment, no residual left-to-right or right-to-left shunting is observed (down).

Figure 9: Repeat aortogram after release of the device showing no significant residual shunting.

closure using Amplatzer Septal Occluders; however, balloon sizing is usually not necessary during PFO closure (Figs. 5 and 6).

Complications during or after transcatheter PFO closure using Amplatzer PFO Occluders are exceptionally rare.

Patent Ductus Arteriosus (PDA)

Isolated persistence of ductus arteriosus (PDA) accounts for approximately 5% of all congenital heart diseases, although it is seen even more frequently in association with complex heart lesions. The ductus arteriosus is a wide muscular blood vessel joining the left pulmonary artery to the aorta.

This connection allows blood to be diverted from the lungs into the aorta during foetal development, since the baby does not breathe until after delivery. The ductus normally closes within 10 days after birth. If it fails to close, PDA occurs and blood continues to flow from the aorta into the pulmonary artery (Fig. 7).

A PDA is a source of left-to-right shunting causing increased pulmonary blood flow. If it is large, it may cause heart failure, failure to thrive and pulmonary arterial hypertension. More common is a small PDA, presenting as a continuous murmur in an asymptomatic patient. These are closed because of the risk of bacterial endarteritis. Percutaneous closure of the patent ductus arteriosus is the treatment of choice (as opposed to surgery), except in very small neonates, especially in premature babies.

Smaller PDAs are treated by transcatheter coil occlusion. The Amplatzer Duct Occluder is a relatively new device designed to improve occlusion rates of moderate to large PDAs (Fig. 8).

This is a self-expanding mushroom-shaped device, with a thin aortic retention disc designed to secure position-

ing in the aortic ampulla.

The device is advanced from the venous side through the long delivery sheath across the PDA in the descending aorta, where the retention disk is opened. The retention disk is snugged against the aortic end of the ampulla. Then, the tubular frame of the prosthesis is deployed into the PDA. With the device still attached to the cable, a descending aortogram is performed in the lateral projection to confirm device position. If malposition occurs, the device can be retracted back inside the delivery sheath. When the proper position is confirmed, the device is released (Fig. 9).

The incidence of complications in this procedure is quite low and includes device embolization, incomplete closure, mild left pulmonary arterial stenosis (in the smallest infants) and very rarely

located entirely within the muscular portion of the interventricular septum and account for 15% of all VSDs. Ventricular septal defects allow left-to-right shunting of blood and pulmonary overcirculation. Patients with large VSDs present early in life with signs and symptoms of congestive heart failure and failure to thrive. These patients undergo surgical repair. Those with moderate-size defects that can be managed medically are candidates for device closure. The presence of left ventricle volume overload on echo, cardiomegaly on chest x-ray or an episode of infective endocarditis are criteria for device closure.

Percutaneus closure has been described of muscular and perimembranous congenital VSDs. Despite being the most common congenital heart defect, transcatheter closure of VSDs is still an uncommon procedure in most tertiary centres, as it is technically more challenging than that of closing an ASD II, PFO or PDA. It requires the formation of a continuous arteriovenous loop for delivery of the device from the venous side. Further, a perimembranous VSD should have a margin of at least 2 mm from the aortic valve before it can be taken up for transcatheter closure using a modified Amplatzer device (Amplatzer
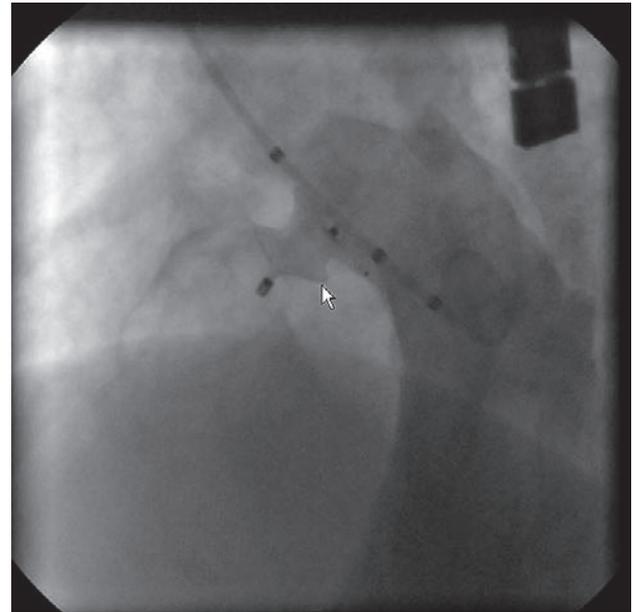
Figure 11: Ventriculogram demonstrating appropriate device position and shape without residual shunt across the defect.

PM VSD Occluder), which has been specially constructed to reduce the risk of any adverse effect on the valves in proximity of the defect – the aortic and tricuspid valves. The first closures of perimembranous VSDs were performed in our centre in autumn 2005, with very encouraging early clinical results (Fig. 11).

Once the device has been implanted, embolism is a rare occurrence and complete closure can be rapidly achieved in most cases. Valvular regurgitation may occur due to impingement of the device against the affected valve. Ventricular arrhythmias during the procedure and conduction disturbances afterwards are possible. Other rare complications include air embolism and pericardial effusion. However, altogether, complications are rare.

Figure 10: Left ventriculogram showing substantial shunting of dye (arrow) through a perimembranous ventricular septal defect.

haemolysis (secondary to incomplete closure).

Ventricular septal defect (VSD)

Ventricular septal defects account for approximately 20% of all forms of congenital heart disease. More than 70% of all VSDs are perimembranous (PM VSD) and are located in the mid-portion of the ventricular septum (Fig. 10).

Muscular defects (Musc VSD) are

Perimembranous VSD occluder

Conclusion

Since the beginning in the year 2000, much clinical experience has been accumulated by our team at the University Medical Center in Ljubljana regarding transcatheter closure of various congenital heart defects using different types of Amplatzer occluders. Along with the obvious advantages in comparison to surgery, we believe that with careful preselection of patients and careful attention to technical details, transcatheter management of congenital heart defects using different types of Amplatzer occluders is safe and effective.