

Current and Novel Devices in Structural Heart Disease

Tobis JM^{1*}, Abudayyeh I²

1 UCLA David Geffen School of Medicine, Division of Cardiology, 10833 Le Conte Ave, Factor Building, Room B-976, Los Angeles, CA 90095, USA

2 Division of Cardiology, Interventional Cardiology, Loma Linda University Health, 11234 Anderson Street, MC 4404, Loma Linda, CA, 92354, USA

* **Corresponding author:** Jonathan Tobis, Professor of Medicine, Director of Interventional Cardiology Research, UCLA David Geffen School of Medicine, Division of Cardiology, 10833 Le Conte Ave, Factor Building, Room B-976, Los Angeles, CA 90095, 310 794-4797, Tel: 3102670384; E-mail: jtobis@mednet.ucla.edu

Abstract

The new field of structural heart disease has gathered significant momentum in recent years. While the bulk of intervention in cardiology still, by far, involve coronary intervention, structural interventions now rival the results of open heart surgery. We review in this paper some of the devices available for treatment of a variety of structural and congenital cardiac conditions as well as acquired valvular abnormalities. Transcatheter treatment offers advantages over surgical intervention in recovery time, improved patient satisfaction, lower procedural risk and avoidance of cardio-pulmonary bypass especially in high-risk patients. With operator experience, the procedural risks also improve compared to open surgical repairs.

Keywords: Structural heart disease, Interventional cardiology, Catheter based therapy

Introduction

The new field of structural heart disease has gathered significant momentum in recent years. While coronary procedures still comprise the bulk of interventions in cardiology, structural interventions have become more common and the results now rival those of open heart surgery.¹ This paper reviews some of the devices available for treatment of a variety of structural cardiac conditions such as adults with congenital defects as well as acquired valvular abnormalities. Transcatheter treatment offers advantages over surgical intervention in recovery time, improved patient satisfaction, lower procedural risk and avoidance of cardio-pulmonary bypass especially in high-risk patients.

Atrial Septal Defects

Atrial septal defects (ASD) are the second most common congenital heart defect occurring in 8 of 1000 live births. Small defects less than 1 cm may not have clinical symptoms so may not require closure. Larger defects with a shunt ratio greater than 1.4 may produce hemodynamic consequences consisting of atrial arrhythmias or right heart failure and should be considered for closure. Only secundum ASDs may be closed by transcatheter devices provided the anatomy is amenable with sufficient rims to hold a device. All other types require open chest surgery. The most common device is the Amplatzer septal occluder (ASO, St. Jude Medical, St. Paul, MN, USA) (Figure I). The device is composed of a self-expanding double disk with a short connecting waist that

acts to center the device within the defect. The device is made from a 0.004-0.008 inch nitinol wire mesh that covers a polyester material to reduce blood flow through the device. Fibrous tissue in growth occurs in a few months to provide a biologic seal. The size of the device is selected by measuring the diameter of the septal defect, usually with a soft sizing balloon. The Amplatzer device is available in sizes of 4 mm to 38 mm in the United States. The edge overhang for the atrial disks ranges from 6 mm to 8 mm. The device is easy to deploy except for very large ASDs and has proven to be reliable with a low risk of complications or failure.² An ASD may also exhibit multiple fenestrations in the inter-atrial septum, which are too small to accommodate an ASO waist. These may require the use of the Amplatzer Multi-Fenestrated Septal Occluder or cribriform device (Figure II). Unlike the ASO, this device has matched atrial disk sizes to ensure maximal coverage of surrounding fenestrations, and a narrow waist to pass through the smaller defects. Two or three devices can be implanted close to each other to cover a wider area or an aneurysmal fenestrated septum. Long-term outcomes and facility of deployment is similar to the ASO. Complications related to the ASO range between 0.6 and 1%, with the worst complication, erosion, occurring in approximately 0.1%.³ These erosions are most common in children with larger defects requiring relatively larger devices. Insufficient retro-aortic or posterior rims, and Septal mal-alignment were found to be the most common factors. The device most commonly responsible for erosions is the 26 mm occluder. This is believed to be due to the heavier wire gauge used in that device construction relative to the device size. The erosion may occur acutely after plant or several months later. Although rare, allergy to Nickel may cause complications including chest pain due to inflammation that requires surgical extraction of the device in about 0.2%.⁴ A second device commonly used to close ASDs is the Gore-Helex septal occluder (Gore and Associates, Inc. Flagstaff USA) (Figure IIIa). The device is composed of a nitinol wire on an ePTFE patch creating a helix with 1.5 turns in the left atrium and 1.5 turns on the right side. The device is not self-centering so a device to defect diameter size of 2:1 is required so that an edge cannot prolapse through the ASD. The Gore-Helex occluder is a good option for smaller defects less than 13 mm in diameter. No erosions or allergic reactions to nickel have been documented with this device. There is a higher likelihood of wire fracture but this has not translated to an adverse clinical outcome or displacement. The new Cardioform Device by WL Gore and Associates was recently released (Figure IV). This device is composed of a 5-wire support frame covered with ePTFE. The device is intended to conform better to septal anatomy while maintaining stronger radial compression and reduced shunting compared to their previous design. This device is quite easy to deliver and release, with good sealing characteristics. The Cardio Seal and STAR flex devices (NMT Medical, Inc. Boston, USA) were previously available for ASD closures. The devices are no longer available due to a high frequency of residual shunts, lower procedural success rates of 85%, and a higher incidence of thrombus forming on the device.⁵

Patent Foramen Ovale

A patent foramen ovale (PFO) is a congenital inter-atrial pathway that persists in 20-30% of the population. In utero, the foramen ovale permits shunting of oxygenated placental blood from the IVC to bypass the fluid-filled fetal lungs, enter the left cardiac side and perfuse the critical organs.⁶ After birth, venous return no longer provides oxygenated blood and the septum primum and septum secundum fuse to become the foramen ovale. Should fusion not occur, the foramen ovale remains patent but is typically asymptomatic. In a minority of patients, symptoms do occur as there is an association of PFO with cryptogenic stroke, migraines with aura, orthodeoxia platypnea, decompression illness, high altitude pulmonary edema, and exacerbation of obstructive sleep apnea.⁷ The ASO and Gore-Helex devices discussed above for closure of an ASD are also used for PFOs

with similar success and low complication rates. These devices, however, are not approved by the FDA for treatment of PFOs and their use is considered off-label in the USA. An Amplatzer device designed specifically for PFO anatomy is available outside the United States.

Patent Ductus Arteriosus

The patent ductus arteriosus (PDA) is a second fetal shunt that carries oxygenated placental blood from the pulmonary artery to the aorta, bypassing the lungs in utero. A PDA is less likely to be present in adults since most PDA shunts are diagnosed in childhood and treated. Prior to closure of a PDA, a full hemodynamic study should be performed to judge suitability of closure and to measure the duct length and diameter by bi-plane angiography during opacification of the duct. Transcatheter closure can be accomplished using embolization coils such as Gianturco coils (Figure V), the Amplatzer duct occluder (Figure VI), or the Nit-Occlud device (Figure VII) (pfm medical ag, Köln, Germany). To deliver the device to the appropriate location in the ductus, the operator advances a wire from the arterial side and enters the PDA through the aorta. The wire is then snared in the pulmonary artery and externalized creating a rail. The device is then advanced across the PDA from the pulmonary artery side over the wire to deploy the occluder device. Two versions of the Amplatzer duct occluder (ADO) exist. The ADO-I has a conical shape with a retention skirt on the arterial side to secure the device. It has a relatively simple deployment technique and high success rate with over 95% of treated patients achieving ductal closure at 6 months.⁸ Sizing selection requires that the smaller end of the device be at least 2 mm larger than the narrowest portion of the PDA measured by angiography. The ADO-II device is a compliant dual articulating disc device with a symmetrical design. It allows the operator to achieve closure from either the aortic or pulmonary artery side as both sides have a retention disc. More attention has to be paid to the waist length on this device compared to the duct to allow the distal disc to expand appropriately. The Nit-Occlude PDA occlusion device has a traumatic biconical configuration and variable stiffness. The larger conical component is more firm and conforms to the aortic side forming a plug with the rest of the coil winding inside the ductal tunnel. A small coil on the pulmonary side anchors the device in place.

Ventricular Septal Defects

A ventricular septal defect (VSD) is an incomplete fusion of the inter-ventricular wall. It can exist in the muscular ventricular septum, the membranous portion close to the atrioventricular node, sub-arterial or supracristal, or as an inlet type VSD.⁹ The most common defect is in the muscular septum, which, if small, usually closes spontaneously in childhood. Membranous VSDs are more common in older children and adults. The defects that may be managed by transcatheter techniques are the perimembranous and muscular VSDs. VSDs that occur post myocardial infarct with rupture of the ventricular septum are associated with a high mortality but may be amenable to percutaneous closure in selected patients. VSDs can be associated with significant sequel including heart failure, pulmonary hypertension, conduction abnormalities, and valvular dysfunction.¹⁰ Transcatheter closure of a VSD can be technically challenging but may produce long term outcomes similar to surgery.¹¹⁻¹² Options for closure include using the Amplatzer series of VSD devices and the Nit-Occlud device. The Amplatzer muscular VSD occluder is a double-disk nitinol wire mesh with polyester patch design similar to the ASD occluder, but a wider waist to fit the thicker muscular septum. There is a thinner membranous VSD device and a post-myocardial infarction device with a longer waist to accommodate damaged muscular tissue. The muscular occluder has symmetric right and left ventricular disks 8 mm larger than the waist. The size of the

device is determined using imaging and possibly a sizing balloon to measure the width of the VSD (Figure VIII). The post-infarct version of the device has a 10 mm long waist to accommodate infarcted myocardial tissue at the rims (Figure IX). The membranous VSD occluder is also a self-expanding, self-centering retrievable double-disk device with a shorter waist. The device is eccentric with a left side aortic rim of 0.5 mm and a left side ventricular rim of 5.5 mm. The ventricular end has a radio-opaque marker to help orient the longer component of the disk towards the ventricular apex. This allows the device to be implanted without interfering with the heart valves (Figure X). A version of the Nit-Occlud device is available for perimembranous VSD closure. This device contains polyester fibers to improve closure. The device is well suited for aneurysmal perimembranous VSDs, and there are no reports of permanent AV-block.¹³ To size the Nit-Occluder, a coil with a distal diameter at least twice the minimal diameter of the VSD on the right ventricular side and 1 to 2 mm greater than the diameter of the VSD at the left ventricular side should be selected.

Aortic Stenosis

Over the past 10 years, transcatheter aortic valve replacement (TAVR) has become a reasonable option for treatment of degenerative aortic stenosis. Standard open-heart surgery or trans-catheter repair substantially changes the natural course and predicted survival of severe aortic stenosis.¹⁴ The Edwards Sapien valve (Edwards Life sciences, Irvine USA) was studied in the PARTNER trial. The original Sapien valve was composed of three leaflets made from bovine pericardial tissue sutured together to a polyethylene terephthalate (PET) skirt on a balloon expandable stainless steel stent frame (Figure XI). It was available in one of 2 diameter sizes: 23 mm or 26 mm. The valve was crimped on a balloon delivery catheter and advanced retrograde through the femoral artery (Figure XII). Alternatively, the valve could be delivered via a trans-apical access through a mini-thoracotomy in the lateral chest or through the distal ascending aorta via a midline mini-thoracotomy. The second generation Edwards valve, the Sapien XT, was a bovine pericardial tissue valve as well, but was sutured on a cobalt chromium stent scaffold (Figure XIII). This stent construction allows the valve to be crimped to a lower profile while maintaining structural and radial support once expanded. The Nova Flex+ delivery catheter is a stretchable sheath with a lower vascular access profile. To diminish the arteriotomy size, the valve is not crimped on the balloon directly during preparation, but is crimped proximal to the balloon. Once the delivery catheter is in the ascending aorta, the balloon is pulled into the valve and is then advanced to the correct position. The Sapien XT was available in three diameter sizes: 23 mm, 26 mm, and 29 mm accommodating aortic annulus areas greater than 490 mm². The delivery sheath for this device includes sizes of 16, 18 and 20 Fr requiring iliac artery diameters of 6, 6.5, and 7 mm respectively. This resulted in lowered vascular complication rates compared to the first generation valves. The incidence of conduction injury remained low at 6%.¹⁴ The newest generation of Tran's catheter valves from Edwards is the Sapien 3 (Figure XIV). This valve was approved in the US in 2015. It is also a bovine pericardial leaflet valve mounted on a modified lower profile cobalt chromium stent frame. An outer skirt helps reduce para-valvular leaks. The redesigned Edwards Commander delivery catheter provides additional dual articulation to help better position the valve coaxially in the outflow tract. Improvements in the design of the catheter and sheath have resulted in a delivery profile of 14 French for the 23 mm and 26 mm valves, and 16 French for the 29 mm valves. These require vessel sizes of 5.5 mm and 6 mm respectively. There is an even lower rate of vascular complications but an increased rate of conduction injury and pacemaker implants. The Core valve is produced by Medtronic (Minneapolis, USA). The valve gained FDA approval in 2014 for patients with severe symptomatic aortic stenosis who are at high risk for surgical intervention. The SurTAVI trial demonstrated the

efficacy of the Core valve compared to surgical intervention. The stroke rates were equivalent at 5.8% and 7% respectively ($p=0.6$). The risk of needing a permanent pacemaker was significantly higher than with the balloon expandable valves at 25 to 30%. The Core valve is made from porcine pericardium on a self-expanding nitinol frame (Figure XV). The thinner tissue of porcine pericardium, compared to bovine, as well as the self-expanding design allowed for a smaller delivery catheter profile of 18 French. The valve is available in sizes ranging from 23-31 mm diameter, which is useful in patients with a larger aortic annulus. The higher incidence of conduction block was due to the longer stent design (55 mm) reaching further down the left ventricular outflow tract and the self-expanding nature of the valve, which exerts pressure on the conducting tissue within the septal wall. Careful valve positioning has resulted in a lower need for permanent pacemaker implantation of 15%.¹⁵ Additionally, the persistent radial pressure of the valve scaffold on the aortic wall theoretically decreases para-valvular leaks over time; however this has not been directly studied. The Core valve does not require rapid pacing to be deployed, but moderate pacing of 90-120 bpm may better stabilize the valve in patients with severe regurgitation or ectopy. St. Jude Medical is evaluating a transcatheter valve, the Portico valve, in the TF CE Trial. This valve consists of bovine pericardial tissue on a self-expanding nitinol stent structure. The stent is shorter than the Core valve reducing the risk for conduction injury. The device also does not require rapid pacing for deployment. The Portico valve is deployed through an 18 French delivery sheath. Currently, only a 23 mm valve is available but a 25 mm valve is being developed (Figure XVI). The direct flow valve (Direct flow Medical, Santa Rosa, CA) is a polymer valve made of bovine pericardium, and an expandable cuff (Figure XVII). Once the valve is across the annulus, the cuff is inflated with a liquid plastic polymer, which hardens to secure the cuff in place. The system is introduced using an 18 F sheath. Smaller trials were conducted in the US and the larger SALUS trial was initiated in May 2014. Currently in the US, only patients with severe symptomatic degenerative aortic stenosis qualify for TAVR. Aortic valve-in-valve procedures where a previously implanted bio-prosthetic valve has degenerated in a high surgical risk patient have recently gained FDA clearance. Trans-catheter valves have also been used to correct valvular dysfunction in the mitral, pulmonic and tricuspid positions.

Mitral Stenosis

Mitral stenosis has decreased in incidence in developed countries but continues to be prevalent in much of the world. The primary cause is rheumatic mitral disease. Other less common causes include infective endocarditis, severe mitral calcification, lupus erythematosus, infiltrative disease, and advanced carcinoid. As the mitral valve area decreases, filling of the left ventricle is impaired and, with higher heart rates, the diastolic filling time is reduced. This produces increased left atrial pressure and pulmonary congestion.¹⁶ The left atrium becomes dilated causing the incidence of atrial fibrillation and thromboembolic to increase substantially. Percutaneous balloon commissurotomy can be performed using a double balloon technique, or with the single Inoue balloon. The Inoue balloon has made mitral commissurotomy a more stable and straightforward minimally invasive procedure (Figure XVIII). The balloon is made of poly-ester micromesh between two latex layers. The balloon is self-centering due to the waist in the middle. It is available in three sizes that cover a range of 24-30 mm in diameter. Careful review of the mitral valve anatomy is required to assess suitability; heavily calcified commissures are unlikely to yield and may rupture. A calibrated syringe helps fill the balloon to the desired diameter in a progressive fashion. A balloon stretching tube is used with the inner tube to stretch the Inoue balloon as it is introduced into the body and across the septum to slenderize it, giving it a lower profile. A curved stylet directs the balloon at the appropriate angle to help cross the stenotic mitral valve

(Figure XIX). When performed by experienced operators, the procedure has a high technical success rate of 89%.

Mitral Regurgitation

Mitral regurgitation (MR) is more common than mitral stenosis and is categorized depending on whether there is a structural abnormality of the valve leaflets or chordae or if the regurgitation is due to valve dysfunction created by LV dilatation. The most common cause of structural MR is valve prolapse due to myxomatous degeneration and chordal stretching.¹⁷ There are several devices that have been developed for percutaneous repair of mitral regurgitation including the MitraClip, Carillon, Cardioband, MONARC, Mitralign, and trans-catheter mitral valve replacement. The MitraClip (Abbott Laboratories, Abbott Park, IL) was developed to percutaneously recreate the Alfieri stitch described in 1991 to treat mitral regurgitation by creating a double orifice for the mitral valve by clipping the middle edges of the anterior and posterior mitral leaflets together. The Everest II trial demonstrated efficacy of the device in high-risk surgical patients and MitraClip has gained widespread use outside the US with approximately 30,000 cases performed (Figure XX). The device is composed of a steerable 24 Fr guide-catheter that is placed from the IVC across the atrial septum. The MitraClip device is made of cobalt chromium with a polyester cover. The delivery system controls two separate MitraClip arms to stabilize then grasp the mitral leaflets. Once an appropriate position is attained, the clip is closed and the leaflets are secured and partially pulled together. The EVEREST II trial demonstrated improved safety in the trans catheter treated group primarily due to lower bleeding rates compared with surgery. High risk patients (STS score >12) with functional mitral regurgitation demonstrated procedural success between 73 and 100% that was sustained at 12 months. The need for mitral valve surgery at one year was low at under 6% with maintenance of NYHA functional class I/II for 75% of patients.¹⁸ The most important component of this procedure is Trans-Esophageal Echo guidance (TEE). Specific views are needed in the hands of an experienced operator to guide the device into the proper position and plane. A second percutaneous approach to treating MR is to simulate a surgical annuloplasty by using the anatomic proximity of the great cardiac vein to the mitral annulus. The Carillon device (Cardiac Dimensions, Inc Kirkland WA) is delivered from the right internal jugular vein through the coronary sinus just proximal to the takeoff of the anterior interventricular vein. A distal anchor is secured and then the atrial tissue is plicated by pulling tension on the device, which simultaneously pulls on the tissue around the mitral annulus. The plication of tissue approximates the anterior and posterior mitral leaflets to decrease the regurgitant flow. There is a risk of circumflex coronary artery compression as branches of this vessel may pass under the great cardiac vein in 27% of hearts. As the device is delivered via the right heart and through the coronary sinus, the procedural risk is low. The Carillon mitral contour system is a coronary sinus implant that consists of a proximal and distal anchor connected by a nitinol shaping ribbon (Figure XXI). The distal anchor is delivered in the great cardiac vein and after tension is applied, the proximal anchor is delivered near the ostium of the coronary sinus. The immediate effect on MR is assessed by TEE and coronary compression is evaluated before release. The tension can then be adjusted or the device retrieved as necessary. The Mitralign (Mitralign, Tewksbury, Massachusetts) is a direct mitral annuloplasty device. This device delivers two pledgette sutures into the opposing posterior commissures of the mitral valve. The sutures are then plicated, tightening the posterior leaflet and reducing the mitral annulus diameter. The guided delivery systems device (Guided Delivery Systems, Santa Clara, California) introduces a catheter into the left ventricle retrograde from the aorta. The catheter is then used to deliver several anchors on the ventricular aspect of the posterior leaflet. The anchors are “cinched” together reducing the valve annular diameter.

This device also shows great promise in treating functional tricuspid regurgitation. The cardioband device is similar in that it delivers a flexible ring onto the atrial side of the valve to “cinch” the posterior leaflet closer. Due to the more complex anatomy of the mitral valve and surrounding structures, a trans-catheter solution for mitral valve replacement is still not mature at this time. If the patient already has a surgically implanted bio-prosthetic valve, the option exists for implanting a currently available device such as the Edwards Sapien valve as a “valve-in-valve” procedure. The previously implanted prosthesis acts as an anchoring ring for the valve stent with good procedural success rates and improvement in outcomes and function.¹⁹

Pulmonic Valve Stenosis and Regurgitation

Improvement in the management of congenital heart disease patients at an early stage has increased survival. Pulmonic valve stenosis may be congenital, acquired, or associated with more complex congenital heart diseases. Balloon valvuloplasty can be used in isolated stenosis with good long-term outcomes [20]. Valve regurgitation often occurs in the same population, especially after prior congenital repair such as the Ross procedure or Tetralogy of Fallot repair. Trans-catheter valves are increasingly used in these cases or where the valve or conduit has degenerated to produce severe pulmonic insufficiency. The melody valve (Medtronic, Minneapolis, MN) is commercially available with indications for pulmonic regurgitation or stenosis and a dysfunctional right ventricle with a conduit size greater than 16 mm. It is a bovine jugular valve on a platinum-iridium stent crimped on a 22 Fr Ensemble catheter delivery system (Figure XXII). The delivery catheter is a balloon-in-balloon system with outer diameters of 18, 20, and 22 mm introduced via the femoral vein. Edwards Life sciences are evaluating the Sapien pulmonic heart valve. This is a bovine pericardial tissue valve on a steel frame similar to the Edwards aortic Sapien valve. Many of these patients have complex anatomy or unusual cardiac orientations so assessment at the time of implant by angiography or pre-procedure CT is required to avoid placing the valves in dilated or deformed conduits, or cause compression of coronary arteries.

Paravalvular Leaks

Paravalvular leaks occur when the annulus of an implanted valve does not fully oppose the annulus around it, which may occur due to disruption or dehiscence of ring sutures or annulus dilation. The leaks may be hemodynamically significant or can be associated with symptomatic hemolysis. Even moderate regurgitation results in worse patient outcomes.²¹ No device is currently approved in the United States and catheter-based interventions are performed off-label. Vascular plugs are most often used for closure. The most common of these are the Amplatzer family of plugs (St. Jude Medical, St. Paul, MN, USA). These plugs are made of a nitinol mesh delivered by a cable through a catheter system. The Amplatzer Vascular Plug (AVP) is a single lobe self-expanding plug available in diameters from 4 mm to 16 mm (Figure XXIII). The AVP-II is multi-segmented with discs on both ends. These stabilize the device in larger leaks and limit flow through the defect reducing time to occlusion (Figure XXIV). It is available in sizes from 3 mm to 22 mm. The AVP-4 is a low-profile bi-lobed device on a flexible delivery wire that allows for easier maneuverability (Figure XXV). This device can be delivered through a 0.035 inch ID (4 Fr) catheter and is available in sizes from 4 to 8 mm. It is also possible to use the ADO-II ductal occluder, described previously, for larger paravalvular leaks. Careful assessment of the defect orientation and size is necessary before and during the procedure. TEE with 3D imaging can be especially helpful to visualize the orientation of the leak relative to the catheter. For crescentic defects, more than one device or a larger device may be needed to cover the area.²² The AVP-III device is

oblong and is beneficial in these crescentric shaped leaks but it is not available in the United States.

Coarctation of the Aorta

Coarctation of the aorta is a congenital condition producing a discrete narrowing of the aorta typically at the ligamentum arteriosum distal to the left subclavian artery. Coarctation of the aorta is associated with bicuspid aortic valves in about 50% of cases and aortopathy (where the aorta dilates in a fusiform aneurysm) in 56% to 88% of those aged 30 and 80 years old respectively.²³ Early repair of the stenosis with an expandable stent improves long term outcomes.²⁴ The procedure has risks as expanding a stent may tear the tunica intima and a rupture is life-threatening. Covered stents are preferable and surgical backup should be available when performing this procedure. The aorta exerts recoil force so a device with high radial strength is necessary. A 14-16 Fr access is obtained in the femoral artery and a long sheath is introduced to the aortic arch past the narrowing. Stents are mounted on a balloon-in-balloon system (BIB, NuMed, Hopkinton, Ky), and (pfm medical ag, Köln, Germany) that allow for a more precise and controlled stent expansion (Figure XXVI). The outer balloons range in size from 8 to 24 mm. Once the stent is in position, the sheath is withdrawn and the stent can be deployed by inflation of the inner then outer balloons. Many stents are available for younger children, but as patients grow, the stents will need to be expanded to a larger diameter of 20-30 mm the most commonly used stents are the Palmaz XL stent, the Palmaz Genesis, Cheatham-Platinum (CP) stent, the IntraStent, and the Atrium Advanta stent.²⁵ The Palmaz XL (Johnson & Johnson, New Brunswick, NJ) is a laser-cut closed-cell stainless steel stent with high radial strength. It is available in lengths of 31, 40 and 50 mm and must be expanded to a minimum diameter of 10 mm limiting its use in smaller children. The Palmaz Genesis stent (Johnson & Johnson, New Brunswick, NJ) is also a stainless steel closed-cell stent with curved segments allowing for improved conformability. This stent is available in lengths of 19,25,29,39 and 59 mm. As this stent is limited at full expansion to 19 mm, it is not optimal for patients who are expected to grow to a normal sized aorta. The CP stent (NuMed, Hopkinton, Ky) is a 0.013" platinum-iridium frame welded at each joint and over brazed with gold. The stent may be expanded from 12.0 mm to 24.0 mm. A covered version of the stent has an expandable sleeve of ePTFE. The IntraStent LD (Covidien, ev3, Plymouth, MN) is an open-cell stainless steel stent with less radial strength but greater flexibility and conformability. The IntraStent LD may be dilated up to 26 mm, and is delivered via a smaller 8 French sheath. The atrium Advantage (Atrium Medical Hudson, NH) series is an encapsulated covered stent where the ePTFE is on both the inside and outside of the stent. It may be dilated up to 22 mm and available with 12, 14, and 16 mm balloons (Figure XXVII).

Peripheral Arteriovenous Fistula

Arteriovenous fistulas (AV fistula) are abnormal communications between arteries and veins. It may occur as a consequence of hereditary hemorrhagic telangiectasia (HHT), due to lack of inhibition of endothelial growth factors, injury, or after surgery such as the Fontan repair. Hemodynamic studies may show persistent elevation of right heart pressures, or reduced peripheral vascular resistance. These AV fistulae tend to be fragile. Pulmonary AV fistulas bypass the lungs delivering de-oxygenated blood to the left circulation.²⁶ Reduction of the pressure causing the AV fistula will resolve peripheral AV fistulae but, along with surgery, percutaneous options are available. Coil embolization is usually successful and relatively safe. For pulmonary AV fistula the operator introduces a Berman catheter and angiograms of the pulmonary artery and veins in the levo-phase are obtained. Once the AV fistula is identified, a catheter is advanced

over a wire to the vessel and the AV fistula is engaged. Coils of the appropriate diameter and length are then introduced to block the abnormal flow.

Left Atrial Appendage Occlusion

Atrial fibrillation increases in prevalence with age and affects more than 10% of people over the age of 80. The lifetime risk of having atrial fibrillation is 25% .27 Atrial fibrillation increases the risk for developing cerebral and non-cerebral embolic events. The CHA2DS2-VAS2 score helps predict the risk of developing emboli for patients with atrial fibrillation who do not have associated mitral stenosis. Approximately 90% of emboli develop in the left atrial appendage due to stagnation of blood flow during atrial fibrillation .28 Trans-catheter devices that occlude the left atrial appendage are attractive as an option to reduce the risk of emboli especially in patients intolerant to anticoagulation therapy or who have an increased risk of bleeding as measured with a high bleeding score (HAS-BLED). The Watchman device (Atritech, Inc., Minneapolis, MN) is a self-expanding nitinol frame with fixation barbs and a permeable polyester fabric that covers the atrial side of the device (Figure XXVIII). The device is available in diameters from 21 to 33 mm. Careful assessment by imaging and angiography should guide selection of the device size and its positioning in the appendage. After a trans-septal puncture, the device is delivered via a 12 Fr catheter into the left atrial appendage. Trans-esophageal imaging is used during the procedure to ensure adequate fixation while covering the entire opening of the appendage and ensuring that residual leaks are eliminated or reduced. The Amplatzer cardiac plug (St. Jude Medical, St. Paul, MN, USA) is also a LAA occluding device that is made from a flexible nitinol wire mesh. The device is softer and more flexible than their ASD occluder series. The device has a self-orienting disc on an articulating neck (Figure XXIX). This allows the distal part of the device to occupy the left atrial appendage lobe for anchoring. The proximal articulating disc then achieves full coverage of the appendage opening and is termed the “garbage can cover” to the LAA (Figure XXX). Device sizing depends on the distal inner-wall lobe and ranges from 16 to 30 mm. The outer disk is 4 mm larger for lobes smaller than 24 mm and 6 mm for larger appendages. The device is not yet approved in the United States. A third device in use is the Lariat suture delivery system (SentreHeart, Redwood, and CA). The device delivers into the pericardial space a 40 mm pre-tied suture loop around the appendage to occlude it from the outside. A wire and catheter is advanced up the IVC and across the septum into the left atrial appendage. The wire contains a magnet at the tip. A pericardial tap is done under the left para-xiphoid area and a second wire with a tip-magnet is advanced along the lateral border of the heart until it meets the first magnet. The suture loop is then advanced along the pericardial wire and manipulated over the appendage until it surrounds the opening of the left atrial appendage. To help in positioning, a balloon is inflated in the appendage. The loop is then tightened to achieve appendage occlusion. Complications of this procedure include a moderate amount of pain, residual pericarditis, residual leaks, and the risks of tearing the soft tissue of the appendage.

References

1. Steinberg DH., Staubach S., Franke J., et al. Defining structural heart disease in the adult patient: current scope, inherent challenges and future directions. *Eur Heart J* 2010; 12: E2-E9.
2. Masura J., Gavora P., Podnar T. Long-term outcome of transcatheter secundum-type atrial septal defect closure using Amplatzer septal occluders. *J Am Coll Cardiol* 2005; 45: 505-507.

3. Peirone AR., Pedra SF., Cardoso Pedra CA. Outcomes after Transcatheter ASD Closure. *Intervent Cardiol Clin* 2013; 2: 39-49.
4. Wertman B., Azarbal B., Riedl M., Tobis J. Adverse Events Associated With Nickel Allergy in Patients Undergoing Percutaneous Atrial Septal Defect or Patent Foramen Ovale Closure. *J Am Coll Cardiol* 2006; 47: 1226-1227.
5. Law MA., Josey J., Justino H., Mullins CE., Ing FF., et al. Long-term follow-up of the STAR Flex device for closure of secundum atrial septal defect. *Catheter Cardiovasc Interv* 2009; 73: 190-195.
6. Hagen PT., Scholz DG., Edwards WD. Incidence and size of patent foramen ovale during the first 10 decades of life: an autopsy study of 965 normal hearts. *Mayo Clin Proc* 1984; 59: 17-20.
7. Tobis J., Shenoda M. Percutaneous treatment of patent foramen ovale and atrial septal defects. *J Am Coll Cardiol* 2012; 60: 1722-1732.
8. Boehm W., Emmel M., Sreeram N. The Amplatzer duct occluder for PDA closure: indications, technique of implantation and clinical outcome. *Images Paediatr Cardiol* 2007; 9: 16-26.
9. Horlick EM., Benson LN., Osten MD. *Structural Heart Disease Interventions*. Lippincott /Williams & Wilkins, pp.2012; 197-206.
10. Hoffman JL., Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol* 2002; 39: 1890-1900.
11. Gabriel HM., Heger M., Innerhofer P., Zehetgruber M., Mundigler G., et al. Long-term outcome of patients with ventricular septal defect considered not to require surgical closure during childhood. *J Am Coll Cardiol* 2002; 39: 1066-1071.
12. Thanopoulos BD., Tsaousis GS., Konstadopoulou GN., et al. Transcatheter Closure of Muscular Septal Ventricular Defects with the Amplatzer Septal Ventricular Occluder: Initial Clinical Application in Children. *J Am Coll Cardiol* 1999; 33: 1395-1399.
13. International Multicenter Clinical Device Investigation on Safety and Effectiveness of the Nit-Occlud® Lê VSD Spiral Coil System for VSD Occlusion" (clinicaltrials.gov identifier NCT00390702)
14. Généreux P., Webb JG., Svensson LG., et al. Vascular complications after transcatheter aortic valve replacement: insights from the PARTNER (Placement of Aortic Transcatheter Valve trial). *J Am Coll Cardiol* 2012; 60: 1043-1052.
15. Lange P., Greif M., Vogel A., Thaumann A., Helbig S., et al. Reduction of pacemaker implantation rates after CoreValve® implantation by moderate predilatation. *EuroIntervention* 2014; 9: 1151-1157.
16. Carabello BA. Modern management of mitral stenosis. *Circulation* 2005; 112: 432-437.
17. Trichon BH., O'Connor CM. Secondary mitral and tricuspid regurgitation accompanying left ventricular systolic dysfunction: is it important, and how is it treated? *Am Heart* 2002; 144: 373.
18. Feldman T., Young A. Percutaneous approaches to valve repair for mitral regurgitation. *J Am Coll Cardiol* 2014; 63: 2057-2068.

19. Webb JG., Wood DA., Ye J., Gurvitch R., Masson JB., et al. Transcatheter valve-in-valve implantation for failed bioprosthetic heart valves. *Circulation* 2010; 121: 1848-1857.
20. McCrindle BW., Kan JS. Long-term results after balloon pulmonary valvuloplasty. *Circulation* 1991; 83: 1915-1922.
21. Krishnaswamy A., Tuzcu EM., Kapadia SR. Percutaneous Paravalvular Leak Closure. *Interventional Cardiology Review* 2013; 9: 44-48.
22. Rihal CS., Sorajja P., Booker JD., et al. Principles of Percutaneous Paravalvular Leak Closure. *J Am Coll Cardiol Intv* 2012; 5: 121-130.
23. Tadros TM., Klein MD., Shapira MM. Ascending Aortic Dilatation Associated With Bicuspid Aortic Valve Pathophysiology, Molecular Biology, and Clinical Implications. *Circulation* 2009; 119: 880-890.
24. Presbitero P., Demarie D., Villani M., Perinetti EA., Riva G., et al. Long term results (15-30 years) of surgical repair of aortic coarctation. *Br Heart J* 1987; 57: 462-467.
25. Garcier JM., Petitcolin V., Filaire M., Mofid R., Azarnouch K., et al. Normal diameter of the thoracic aorta in adults: a magnetic resonance imaging study. *Surg Radiol Anat* 2003; 25: 322-329.
26. Kraemer N., Krombach GA. Images in clinical medicine. Pulmonary arteriovenous fistula. *N Engl J Med* 2009; 360: 1769.
27. Mark Estes NA III., Halperin JL., Calkins H., et al. ACC physician's consortium 2008 Clinical performance measure for adults with non-valvular atrial fibrillation and atrial flutter. A report of the American College of Cardiology. *J Am Coll Cardiol* 2008; 51: 865-884.
28. Abudayyeh I., Passen E., Nicholson D., et al. Fractional Shortening of the Left Atrial Appendage by Multi-detector Computed Tomography. *SCCT* 2011.