Transcatheter Interventions in Adult Congenital Heart Disease

Jamil A. Aboulhosn, MD[a,*], Ziyad M. Hijazi, MD, MPH[b,c]

KEYWORDS

• Adult congenital heart disease • Transcatheter intervention • Pulmonary valve replacement
• VSD closure • ASD closure • Valvuloplasty

KEY POINTS

- Transcatheter interventions are replacing traditional surgical procedures for many types of native and operated congenital heart disease.
- Transcatheter interventions often require fluoroscopic and echocardiographic guidance.
- Transcatheter pulmonary valve replacement is now considered standard of care in patients with failing bioprosthetic valves and conduits.
- New self-expanding devices are undergoing human trials and will allow for transcatheter valve replacement of patients with large native right ventricular outflow tracts and pulmonary regurgitation.

BACKGROUND

The past 3 decades have witnessed an exponential growth of transcatheter interventions for congenital heart disease (CHD). Nowhere has this been more evident than in the adult CHD population, because survival into adulthood is now the norm for most forms of CHD.[1,2] In 1976 King and Mills published the first report of transcatheter device closure of an atrial septal defect (ASD).[3] Since that time, improvements in device design, catheterization technology, and procedural techniques have brought interventional cardiology to the forefront as a therapeutic intervention that may delay or obviate the need for surgery in CHD. Advances in noninvasive cardiovascular imaging have made diagnostic cardiac catheterization in a shrinking pool of patients. Transthoracic echocardiography with Doppler is now the noninvasive imaging work horse for congenital and structural cardiology and is cost effective and widely available. Cross-sectional imaging modalities such as computed tomography scans and MRI provide 3-dimensional volumetric data that are invaluable in the assessment of anatomy and function, especially in those with complex anatomy. The combination of echocardiography and cross-sectional imaging provides a powerful noninvasive armamentarium that is capable of accurately assessing most anatomic and physiologic types of CHD thus relegating diagnostic catheterization to a small subset of patients, typically those with single ventricle physiology, pulmonary hypertension, or those in whom noninvasive imaging results in confusing or contradictory findings. Because most hemodynamic determinations can be made by Doppler echocardiography and anatomic determinations can be made by computed tomography scans or MRI, diagnostic catheterizations are

[a] Ahmanson/UCLA Adult Congenital Heart Disease Center, Ronald Reagan Medical Center at UCLA, 635 Charles Young Drive South, Los Angeles, CA 90095, USA; [b] Sidra Heart Center, Sidra Medicine, Al Gharrafa Street, Ar-Rayyan, Doha, Qatar; [c] Weill Cornell Medicine, Cornell University, 525 East 68th Street, New York, NY 10065, USA
* Corresponding author.
E-mail address: jaboulhosn@mednet.ucla.edu

Cardiol Clin 38 (2020) 403–416
https://doi.org/10.1016/j.ccl.2020.04.005

indicated for determination of hemodynamics that cannot be obtained by other modalities. Diagnostic catheterizations continue to be the gold standard for the evaluation of pulmonary pressures, flows, and resistance, especially in single ventricle patients before and after cavopulmonary shunts.

Interventional catheterization can be therapeutic, reparative, or palliative and is becoming an increasingly popular alternative to cardiac surgery. Further, it can be used to complement and enhance surgical results. Interventional catheterization has largely replaced surgery as the treatment of choice for a number of congenital cardiovascular conditions, including ostium secundum ASD, coarctation of the aorta, patent ductus arteriosus, pulmonary artery or valve stenosis, certain types of ventricular septal defects (VSDs), and conduit or bioprosthetic valve dysfunction. Various devices are under clinical trials for the replacement of pulmonic valves in dysfunctional native right ventricular (RV) outflow tracts. These techniques have shown promise and are expected to enter mainstream clinical practice within the next few years. New procedures and devices are continually evolving. This article details a variety of currently available procedures for adult patients with CHD, including the indications for their use, potential risks, and clinical outcomes.

INTERVENTIONS FOR SEMILUNAR VALVE AND VENTRICULAR OUTFLOW TRACT OBSTRUCTIONS

Right Ventricular Outflow Tract Obstruction

Obstruction of the RV outflow tract can occur at multiple levels, namely, subinfundibular, infundibular, valvular, or supravalvular. Although infundibular obstruction can be effectively relieved in younger patients with stenting, subpulmonic stenosis is typically a surgical problem. Although valvar pulmonic stenosis is very amenable to balloon dilation, conduit stenosis, bioprosthetic pulmonary valve dysfunction, and main and branch pulmonary artery stenoses can all benefit from treatment via transcatheter techniques.

PULMONARY VALVE STENOSIS

Balloon valvuloplasty is the treatment of choice for isolated pulmonary valve stenosis if the valve is mobile and doming. The original catheter based technique was initially described by Rubio and Limon-Lason in 1956.[4] The currently used percutaneous static balloon valvuloplasty technique was first reported by Kan and colleagues[5] in 1982. This procedure has proven safe and efficacious over the past 2 decades, making balloon valvuloplasty the treatment of choice for valvular pulmonary stenosis.[6–8] Although the presence of a dysplastic or heavily calcified pulmonary valve (especially with supravalvular narrowing) is associated with less ideal outcomes with this technique,[6] these patients remain as candidates for pulmonary valvuloplasty.

Nearly 15% of patients with pulmonary stenosis have dysplastic valves. These valves can be distinguished from the mobile doming cases by the absence of a pulmonary ejection sound on physical examination or by noninvasive imaging modalities such as echocardiography or MRI.

Infundibular or subinfundibular RV outflow tract obstruction from hypertrophied myocardium is not amenable to balloon angioplasty. Transcatheter stent deployment has shown limited efficacy for relief of muscular subpulmonary stenosis. In some patients with pulmonary valvular and subvalvular obstruction, relief of the valvular stenosis is often accompanied by gradual regression of the subvalvular hypertrophy and a decrease in the degree of obstruction.[9] In the absence of valvar pulmonary stenosis, medical management with beta-blockers can be attempted; however, surgical relief is frequently needed.

Transcatheter balloon valvuloplasty is the treatment of choice in symptomatic patients with isolated mobile pulmonary valve stenosis if the Doppler-estimated peak instantaneous gradient is 36 mm Hg or greater or in asymptomatic patients with more severe stenosis (gradient $\geq$64 mm Hg).[8,10,11] Peak gradients of less than 36 mm Hg typically do not increase with age, and survival is not curtailed; accordingly, there is no indication for intervention.[11] The technique for pulmonary valvuloplasty has changed little since early reports; however, technological advancements resulting in low-profile catheters have decreased the likelihood of vascular entry site injury. The measurement of the pulmonary valve annulus diameter is an important step before proceeding with balloon selection. The desired balloon/annulus ratio is 1.2 to 1.4.[12] Balloon/annulus ratios that exceed 1.5 risk rupturing the pulmonary valve annulus.[13] If the pulmonary annulus is too large for dilatation with a single balloon, 2 balloons can be used. When using 2 balloons, the combined diameters of the 2 balloons ideally should be 1.2 times the optimal diameter of a single balloon.[14] Two balloons have the advantage of less trauma at the venous access site, success with larger diameter pulmonary valves, and potential decompression of the right ventricle

during valvuloplasty through gaps between the balloons.

Published reports of short- and long-term outcomes have been favorable.[6,7,15] In a study of 25 consecutive adolescents and adults, Sharieff and colleagues[15] achieved procedural success in 80% with an immediate decrease in peak gradient from a mean of 94 mm Hg to 34 mm Hg. The peak gradient decreased further over 3 years to a mean of 19 mm Hg, mainly owing to regression of infundibular hypertrophy. The VACA registry investigators gathered follow-up data on 533 patients who underwent balloon pulmonary valvuloplasty.[7] Over 8 years of follow-up, 23% of patients had a suboptimal outcome, as judged by either a residual peak systolic gradient of 36 mm Hg or greater or the need for further transcatheter or surgical intervention. Predictors of suboptimal outcome included elevated immediate postprocedure gradient (odds ratio, 1.32 per 10 mm Hg increase), a lower ratio of balloon to annulus diameter, and a dysplastic valve. Restenosis is rare. Pulmonary regurgitation is common and is hemodynamically well-tolerated.[16] Compared with surgical valvotomy, transcatheter balloon valvuloplasty results in lower mortality and morbidity and hence is now considered the standard of care with surgery relegated to those with failed transcatheter interventions or incompatible anatomy.[8,17] Although the residual systolic gradients are higher with balloon valvuloplasty versus surgical valvotomy, there is less regurgitation.

PULMONARY ARTERY STENOSIS

Supravalvular pulmonary artery stenosis may occur in isolation, but is most often associated with other lesions such as tetralogy of Fallot. Branch pulmonary artery stenosis often occurs after surgical shunt placement. Native pulmonary artery stenosis can be found in patients with William's, Noonan's, and Alagille's syndromes, or in patients exposed to rubella infection in utero. The indications for transcatheter or surgical intervention in pulmonary artery stenosis include lesions that result in elevation of RV systolic pressure or those that result in relative underperfusion of a lung segment.[18] Surgical repair of pulmonary artery stenosis is possible, but the results are frequently suboptimal, particularly if the lesions are peripheral. Lock and colleagues[19] first described a percutaneous static balloon angioplasty technique for the treatment of peripheral pulmonary artery stenosis in 1983. An adequate result depends on the use of sufficiently large high-pressure balloons that tear the vascular intima and part of the media, leaving a slim safety margin for this procedure.[20] Elastic recoil of the angioplastied segment is common with balloon angioplasty alone, prompting the use of stents.[21] The success rate of transcatheter intervention for peripheral pulmonary stenosis is increased from 70% with balloon angioplasty alone to 90% with the use of stents.[20,21] Cutting balloons are more effective than static balloons in relieving peripheral stenoses that cannot be stented.[22] In general, the first and second arcade branches of the pulmonary arteries are usually effectively treated with stents. Diffuse peripheral pulmonary stenosis can be a very difficult problem for which there is no surgical option (short of lung transplantation). Patients with multiple distal stenosis often require multiple balloon dilations. In some patients, thorough and aggressive dilation of these stenoses can decrease the RV pressure; however, care must be taken to avoid rupture of the distal pulmonary vasculature.

PULMONARY VALVE REPLACEMENT

The single most common application of transcatheter valve replacement in adult CHD is in prosthetic pulmonary valve dysfunction, RV to pulmonary artery conduit failure or the dysfunctional RV outflow tract. The vast majority of adult patients with CHD requiring transcatheter pulmonary valve replacement (TCPVR) are those with repaired tetralogy of Fallot. Other etiologies include congenital pulmonary valve stenosis that has been surgically intervened upon with resultant pulmonary regurgitation, those with repaired truncus arteriosus with subsequently dysfunctional RV to pulmonary artery conduits, and those with congenital aortic valve pathology that have undergone the Ross operation and subsequently developed conduit dysfunction. Among those with tetralogy of Fallot, surgical intervention in infancy is typically performed to patch the VSD and to relieve RV outflow tract obstruction. In those with pulmonary atresia, placement of a RV to pulmonary artery conduit is typical, whereas among those with pulmonary valve stenosis and hypoplastic annulus, surgical relief of the obstruction is often achieved by dividing the annulus of the pulmonary valve and patch augmentation of the RV outflow tract. In past decades, transannular patch augmentation was widely used and therefore this surgical variant is commonly encountered in the adult CHD population, resulting in predominant pulmonary regurgitation and large and often aneurysmal RV outflow tracts. In contrast, patients who have undergone surgical conduit placement (typically using a valved aortic or pulmonic homograft) present with predominant conduit

stenosis or mixed stenosis and regurgitation. Adult patients with CHD born with pulmonary atresia have typically undergone multiple surgical procedures for the placement or replacement of conduits and many have eventually undergone bioprosthetic pulmonary valve replacement. The number of surgical procedures performed correlates with an increase in arrhythmia risk and risk of heart failure.[23] Although the indications and timing of PVR, be it surgical or transcatheter, remain controversial, it is widely accepted that those with severe stenosis and exercise limitation will benefit with subsequent improvement in exercise capacity.[8,24] In adult patients with CHD with predominant pulmonary regurgitation, the 2018 American College of Cardiology/American Heart Association adult CHD guidelines recommend consideration of valve replacement in symptomatic patients or those with evidence of RV or left ventricular systolic dysfunction, severe RV enlargement with an indexed RV end-diastolic volume of greater than 160 mL/m^2, or an indexed RV end-systolic volume of greater than 80 mL/m^2 and those with decreases in exercise capacity.[8]

Homograft conduits, either aortic or pulmonary, have been used extensively since the 1960s and are often present in adult patients with CHD. Progressive dysfunction occurs in the majority of homografts and most require replacement within 15 years from implantation, sooner if implanted in a young child[25] (**Fig. 1**). Heavily calcified aortic homografts are prone to dissection and rupture if aggressively dilated with high-pressure balloons or to beyond their original implantation size and therefore covered stent platforms are often used to decrease the risk of uncontained rupture. Pulmonary homografts typically are not as extensively calcified, but do have a tendency to dilate with resultant regurgitation. Pulmonary homografts can often be dilated to a slightly larger diameter than the original implant diameter if deemed imperative. Infective endocarditis is a serious concern and may have occurred in up to 10% of patients with dysfunctional homografts being considered for intervention; a prior history of endocarditis, immunocompromise, and residual stenosis after TCPVR are risk factor for endocarditis after TCPVR.[26–29] Surgical placement of stented bioprosthetic valves is widely used in adult patients with CHD with dysfunctional conduits and native RV outflow tracts. As with conduits, bioprosthetic valves have a finite life span and the majority require replacement within 15 years of implantation. A dysfunctional bioprosthetic valve provides an ideal landing zone for TCPVR, often referred to as a valve in valve procedure and is associated with excellent outcomes[30,31] (**Fig. 2**). Balloon-expandable transcatheter valves are now widely used to replace dysfunctional atrioventricular bioprosthesis with high success rates and minimal complications (**Fig. 3**). In the presence of

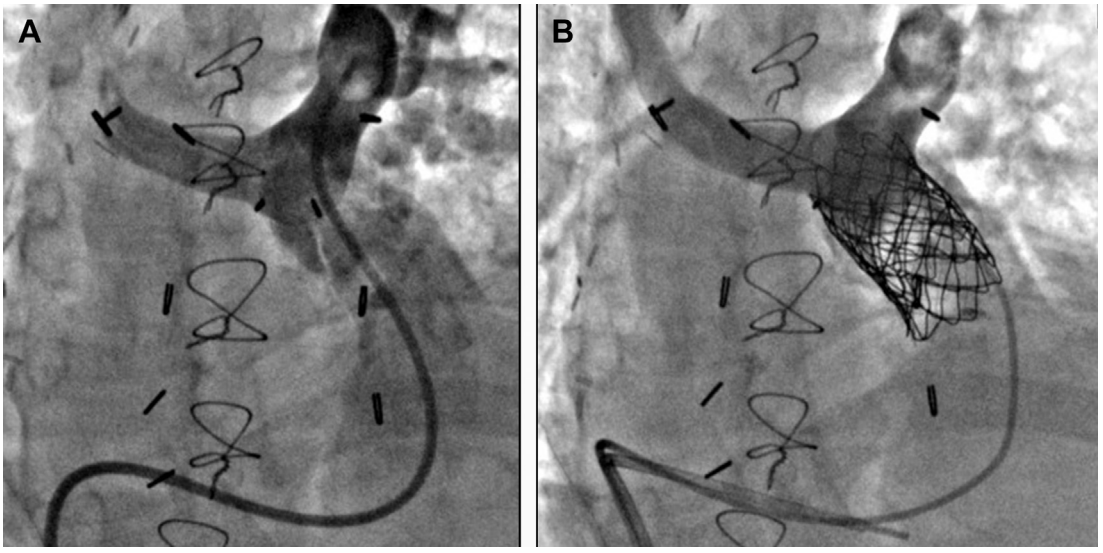

Fig. 1. A 28-year old patient with pulmonary atresia and major aortopulmonary collateral arteries status post bilateral unifocalization and RV to pulmonary artery aortic homograft with progressive stenosis and regurgitation. (*A*) Left pulmonary artery angiogram demonstrating severe pulmonary regurgitation with flow reversal to the right ventricle. (*B*) Status post covered stent placement followed by Melody valve placement with resolution of pulmonary regurgitation.

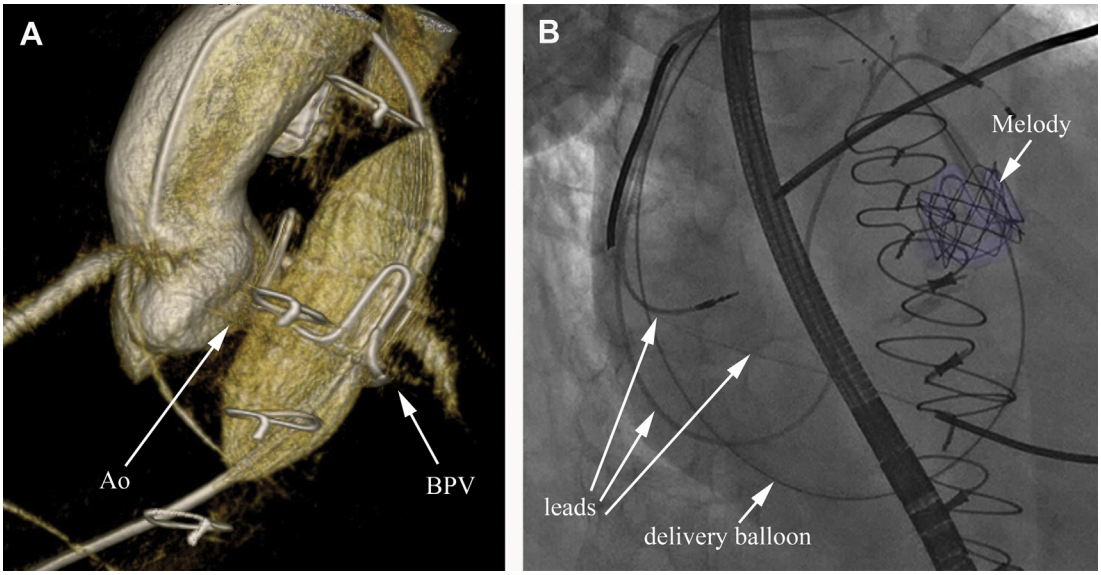

Fig. 2. (*A*) Three-dimensional (3D) rotational aortic angiography with 3D reconstruction during high pressure balloon dilation of a bioprosthetic pulmonary valve (BPV). Note the distortion of the aortic root (Ao). (*B*) Fluoroscopic image post Melody valve placement from a jugular venous approach in a 68 year old man with repaired tetralogy of Fallot, interrupted inferior vena cava, advanced left ventricular failure with biventricular resynchronization and multiple intracardiac leads.

undersized bioprosthetic valves that were placed in childhood, a valve in valve procedure can still be performed by increasing the size of the bioprosthetic valve with the use of high pressure balloons to fracture the valve ring.[32]

In the current era of TCPVR, the most widely used valves in adult CHD in the United States are the Melody valve (Medtronic Minneapolis, MN) and Edwards Sapien Valve (Edwards Lifesciences, Irvine, CA). The Melody valve is a bovine jugular vein cuff and valve sewn onto a platinum iridium stent frame. The valve sizes range from 18 to 22 mm but the valve functions well with a greater range of implant diameters (12–24 mm). The Edwards Sapien valve is now in its third generation; the first generation is no longer available commercially. The second-generation Sapien XT is approved by the US Food and Drug Administration for use in dysfunctional conduits and a prospective trial was just concluded assessing the Sapien S3 in both conduits and bioprosthetic valves (COMPASSION S3 Clinical Trial NCT02744677). Both the XT and S3 are made of bovine pericardial tissue hand sewn onto a cobalt chromium stent platform with the addition of an expanded polytetrafluoroethylene cuff or skirt on the S3 model around the base. The Sapien valves range in size from 20 to 29 mm and are deployed through an expandable sheath design. The larger diameter of the Sapien valves allow for the treatment of conduits, bioprosthesis, and native RV outflow tracts that exceed 24 mm in diameter and therefore cannot be treated with the Melody valve. Both valve platforms are associated with excellent short- and intermediate-term outcomes.[27,33–35] Stent fracture has been noted to be a problem with the Melody valve, especially when prestenting is not performed within conduits or native RV outflow tracts, prestenting does not seem to be necessary in bioprosthetic valves given the presence of a metallic or plastic ring within which the stent platform can be protected from compressive forces.[36] The Sapien valve's cobalt chromium stent frame is significantly more durable, able to withstand high compressive forces, and not prone to fracture; therefore, prestenting before Sapien valve implantation does not seem to be necessary.[37,38] It is imperative to evaluate for coronary arterial compression before valve implantation because coronary compression can occur in approximately 6% of patients.[39] High-risk substrates for coronary artery compression include those with anomalous coronary arterial anatomy and patients with surgically reimplanted coronary arteries.

The treatment of large diameter (>30 mm) native RV outflow tracts is especially challenging, given that the largest commercially available balloon expandable TCPVR platforms are the 29-mm Sapien XT or Sapien 3 valves. The 29-mm Sapien 3 valve can be expanded beyond its nominal diameter by overinflation, with additional volume with

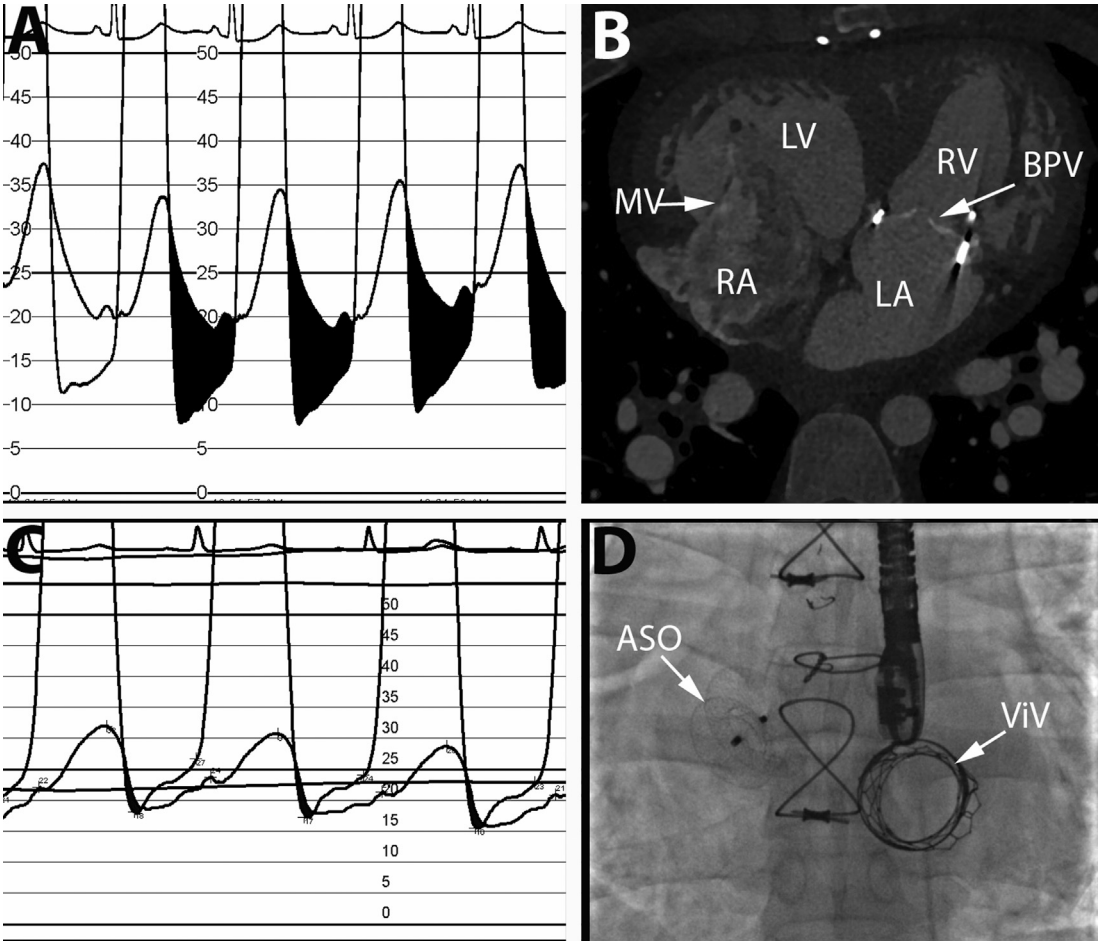

Fig. 3. A 33-year-old with congenitally corrected transposition of the great arteries status post surgical left atrioventricular valve replacement with a bioprosthetic bovine valve (BPV) that has become severely stenotic. (*A*) Simultaneous left atrial (LA) and RV pressures with a mean diastolic gradient of 9 mm Hg. (*B*) Electrocardiogram-gated computed tomography angiogram demonstrating decreased opening of a thickened and partially calcified BPV. In contrast, note the wide opening of the mitral valve between the right atrium (RA) and the subpulmonic left ventricle. (*C*) After trans-septal transcatheter placement of a 29-mm Sapien 3, there is no longer a resting mean diastolic gradient across the valve. (*D*) Fluorscopic en-face view of the Sapien 3 valve within the dysfunctional BPV (valve in valve procedure [ViV]). The large fenestration created within the atrial septum by passage of a 26F sheath was closed with an Amplatzer septal occluder (ASO). MV, mitral valve.

an eventual maximal outer diameter of 31 mm. Hybrid surgical plication of the pulmonary artery can be considered via a sternotomy or thoracotomy to establish a landing zone for TCPVR.[40] The Venus P Valve (Venus Medtech, Hangzhuo, China) and the Harmony valve (Medtronic), are self-expanding covered hourglass-shaped RV outflow tract reducer platforms with the valve in the central waist, both valves are currently in clinical trials. Edwards Life Sciences has developed a self-expanding RV outflow tract reducer that does not house a valve platform, the Alterra adaptive RV outflow tract reducer. The Sapien 3 29 mm valve is subsequently implanted (during same procedure or during a separate procedure) within the Alterra RV outflow tract reducer. This system is currently in clinical trial testing (**Fig. 4**).

Coarctation of the Aorta

Both native and recurrent coarctations of the aorta in adult patients with CHD are usually discrete narrowings beyond the subclavian artery; however, more diffuse forms of the disease may also be encountered, including the presence of a hypoplastic arch or a gothic angulation of the arch. A significant coarctation is defined as having a resting peak to peak gradient of 20 mm Hg or greater.[41] However, significant arterial collaterals may result in unimpressive gradients in those

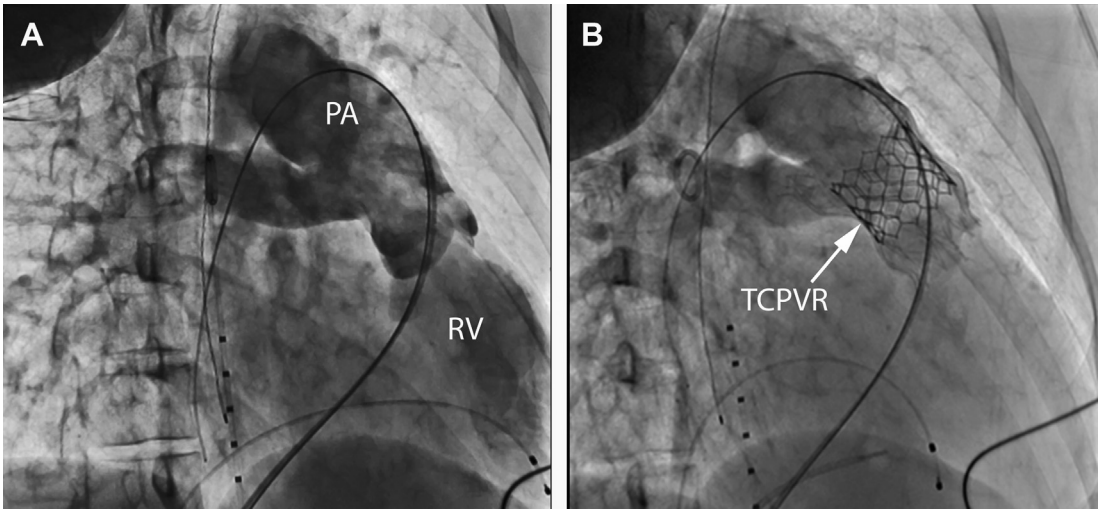

Fig. 4. (*A*) Severe pulmonary regurgitation in a patient with native RV outflow tract. Note the reflux of contrast from the pulmonary artery to the right ventricle (RV). (*B*) After placement of the Alterra adaptive stent with subsequent TCPVR with a 29-mm Sapien 3.

with significant aortic narrowing, additionally, those with poor ventricular systolic function may have lower gradients.

Transcatheter balloon angioplasty of aortic coarctation was initially performed in 1982 by Singer and colleagues.[42] This treatment has been applied to discrete native and recurrent narrowing of the descending aorta with a high immediate success rate; however, aortic wall complications occurred in 23% of patients and reobstruction occurred in 19%.[43–46] An alternative treatment option is stent implantation, first reported by Suarez de Lezo and colleagues[47] in 1995. Open and closed cell design stents and covered stent platforms are now widely used in preference to balloon angioplasty alone (**Fig. 5**). The immediate results are excellent with a low risk of complications (<5%) and a low risk of aortic wall injury (3.1%), the intermediate reobstruction rate is 15%; however, the majority of these are mild restenosis.[43] Intermediate results demonstrate the 3-year freedom from reintervention ranges from 73% to 88% and the rate of aneurysm formation is relatively low.[48,49] The availability of

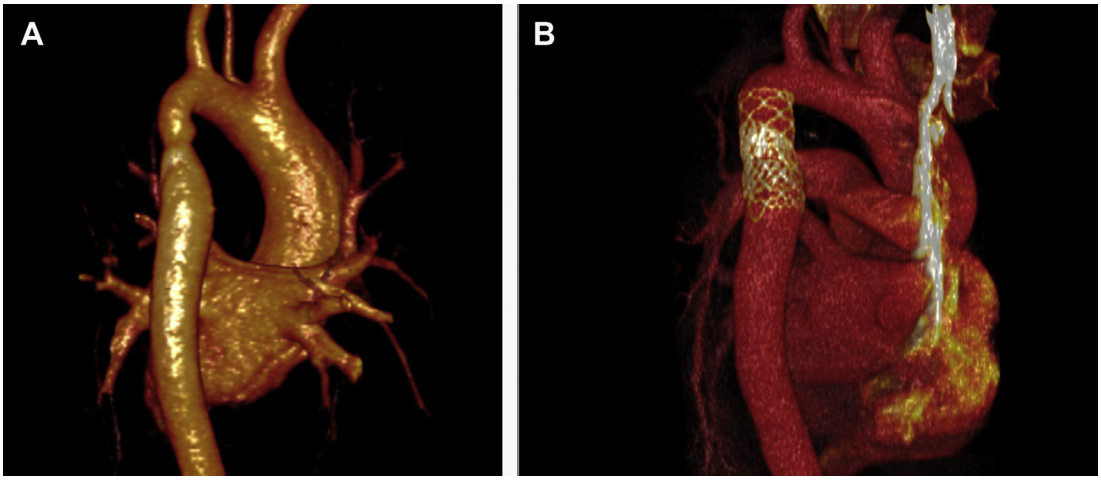

Fig. 5. (*A*) MRI angiography with 3-dimensional volume rendering, right posterior projection, demonstrating severe recurrent coarctation in a patient that had undergone surgical end-to-end repair in infancy. (*B*) Computed tomography angiography, similar projection, after covered Cheatham platinum stent placement and EV-3 Mega LD open cell uncovered stent placement with complete resolution of aortic narrowing.

covered balloon and self-expanding stent platforms may further improve the safety profile of stents and allow for exclusion of aneurysms.[50]

OCCLUSION DEVICES

Patent Ductus Arteriosus

The ductus arteriosus, a communication between the aorta and pulmonary artery, may persist in a wide variety of sizes and forms.[51] PDA closure is indicated in patients with left-to-right shunting who are symptomatic of heart failure or asymptomatic patients with left heart enlargement.[41,52] In adults (and children) with PDA, the outcomes after closure have been very good, including in select patients with moderately increased pulmonary artery pressure and resistance.[53] The Eisenmenger syndrome remains a contraindication to closure. Some authors have advocated the use of temporary balloon occlusion to demonstrate the hemodynamic tolerability of duct closure.[54] Reactivity of the pulmonary vascular bed to pulmonary vasodilating agents and/or a decrease in pulmonary artery pressure and resistance with test balloon occlusion portend a positive outcome with device occlusion. Indications for closure of a small PDA without significant shunting are not well-established and the closure of the silent ductus remains a source of debate. Endarteritis is a rare complication in the current era, but has been reported.[55,56] The closure of all PDAs can be accomplished via catheterization with minimal morbidity and a high rate of success.[57] Currently available devices include coils which are used for closure of small sized PDAs and multiple occlusion devices, including the Amplatzer duct occlude (approved in the United States), Occlutech PDA device, and a variety of Chinese Amplatzer like devices not approved in the United States[58–62] (see **Fig. 7**). The Nit-Occlud device (pfm Medical, Cologne, Germany) is a coil-type device with a controlled delivery system that is the successor to the hourglass-shaped nitinol coil.

Patients with medial aortic wall abnormalities, as in Marfan syndrome and older patients with calcified PDA, should be approached cautiously. PDA occlusion may be performed with aortic covered stent placement.[63] Patients with PDA and pulmonary hypertension but acceptable pulmonary vascular resistance (<6 Woods units) can be closed using an Amplatzer Muscular VSD device to decrease risk of device embolization.

Atrial Septal Defects

ASD is one of the most commonly encountered congenital heart malformations in the adult. The treatment of secundum type ASD has changed drastically over the last 2 decades. Surgical closure had been the preferred treatment since the inception of cardiopulmonary bypass. Although the operative outcomes are excellent, surgical closure carries the risk of complications.[64] Transcatheter device closure has now supplanted surgery as the standard therapy for most secundum ASDs. Accurate defect sizing, determination of adequate rims, and the exclusion of associated cardiac anomalies are the cornerstones to successful percutaneous defect closure.[65] Transthoracic and transesophageal echocardiography are widely used to screen defects for closure and rule out partial anomalous pulmonary venous connections.[66–68] Intracardiac echocardiography has also been advocated as a method for both selection of septal occluder size and for guidance during transcatheter closure.[69,70]

Survival into adulthood is the rule; however, life expectancy is not normal in the patient with an unrepaired defect, with mortality increasing by 6% per year after age 40.[71,72] Progressive dyspnea on exertion and palpitations frequently present in adulthood owing to progressive right heart dilation and dysfunction, pulmonary hypertension, tricuspid regurgitation, and atrial arrhythmias. The degree of left-to-right shunt increases with age as left ventricular compliance decreases and systemic arterial resistance increases. Indications for ASD closure include the presence of significant shunting ad defined by calculating a Qp:Qs of 1.5:1.0 or greater and/or the presence of right heart enlargement on transthoracic echocardiography in the presence of normal or low pulmonary vascular resistance.[41] Defect closure in older adults seems to be beneficial; however, the presence of left ventricular diastolic dysfunction may result in increased left atrial pressure after device closure of ASD. Therefore, careful hemodynamic assessment of the left atrial pressure before closure and during temporary balloon occlusion of the defect is of paramount importance in such patients. The use of fenestrated ASD devices may help in such patients.[73] The exclusion of patients with severe pulmonary hypertension may be obviated by pulmonary artery vasodilator therapy that may decrease pulmonary arterial pressure and resistance, permitting shunt closure in these patients.[74,75]

Advancements in device design and catheterization technology have led to the availability of a variety of transcatheter occlusion devices.[76,77] Transcatheter device closure compares favorably with surgical closure in terms of efficacy and is associated with shorter hospital stays and fewer postprocedural complications.[78] Although the details of the procedure vary depending on the

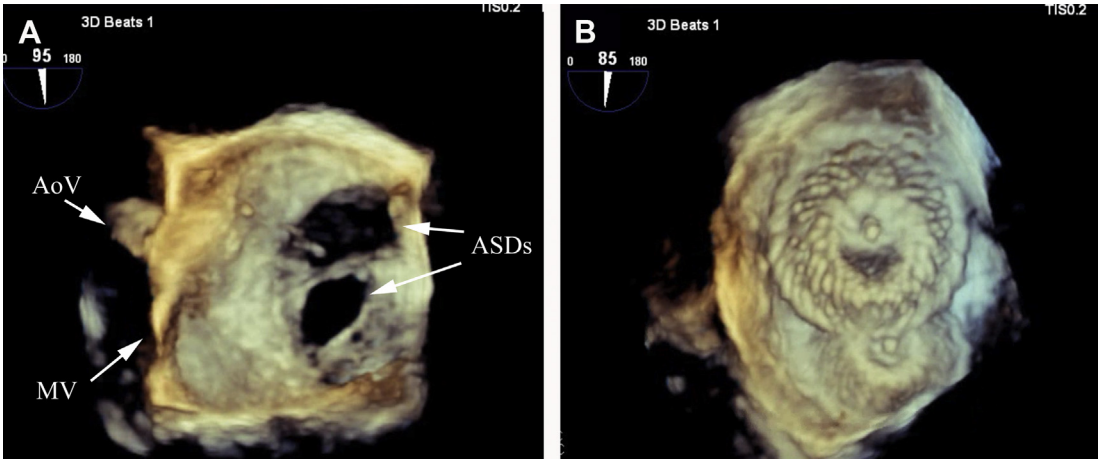

Fig. 6. (*A*) Three-dimensional transesophageal echocardiogram, left atrial view demonstrating 2 large ostium secundum type ASDs, the aortic valve (AoV) and the mitral valve (MV) are labeled for orientation. (*B*) After closure with 2 Amplatzer septal occlude devices (ASD).

device used, in general the procedure is performed under fluoroscopic and transesophageal or intracardiac echocardiographic guidance. Published results vary with each device, but in general, complete closure is achieved in 60% to 90% of cases within hours of deployment, with 80% to 100% closure over 1 year of follow-up.[79] The Amplatzer septal occluder (AGA Medical, Plymouth, MN) remains the most widely used ASD closure device in the United States (**Fig. 6**). The long-term outcomes of device closure using the Amplatzer septal occluder are excellent.[80] Amplatzer septal occluders ranging from 4 to 38 mm have been approved by the US Food and Drug Administration; a 40-mm device is available outside the United States and has been successfully used but there is an increased risk of device embolization.[81] All device types risk rare short-term complications of embolization, thrombus formation, aortic root perforation, pericardial effusion, and dysrhythmias. A variety of devices from W. L. Gore & Associates (Flagstaff, AZ), including the Helex septal occluder, the Cardioform septal occluder, and most recently the Gore Cardioform ASD occluder, can be used for effective ASD closure.[82] Sporadic cases of late erosions have been reported with both the Amplatzer and Gore devices.[83,84] Patients who had received Amplatzer septal occluders with perforation were more likely to have a deficient anterior–superior rim and had received larger device to unstretched defect diameter.[84]

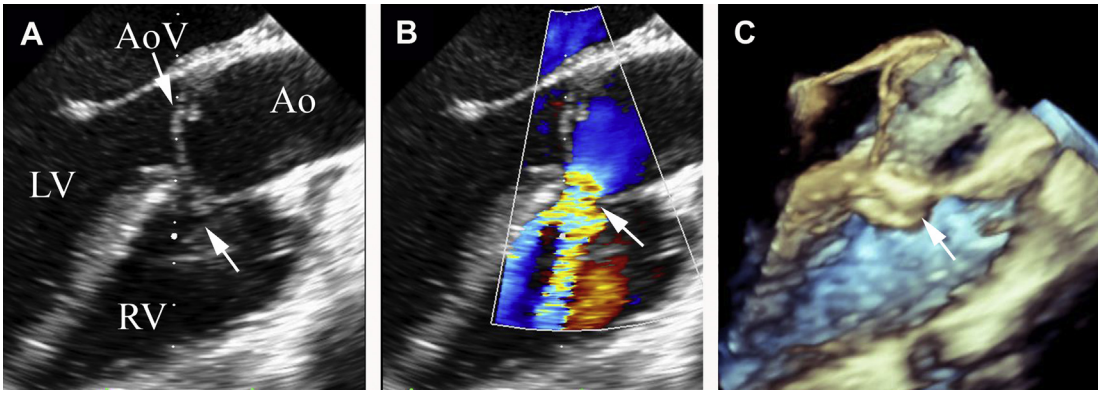

Fig. 7. (*A*) Transesophageal 2-dimensional echocardiography long axis view demonstrating a communication between the aorta (Ao) and the right ventricle (RV) (*white arrow*) above the aortic valve (AoV) via a prolapsing aortic root sinus. (*B*) Color Doppler image demonstrating flow from the Ao to the RV via the ruptured right coronary sinus. (*C*) Three-dimensional transesophageal echocardiography after device closure of the ruptured sinus of Valsalva with an Amplatzer muscular VSD device (*white arrow*).

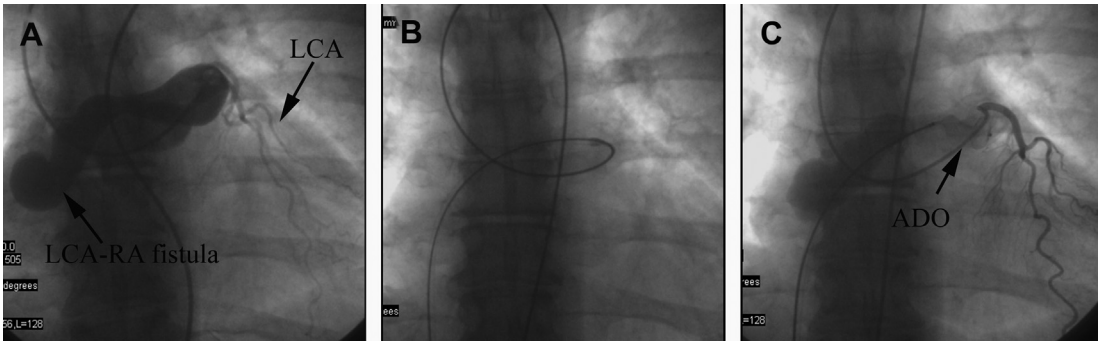

Fig. 8. (*A*) Coronary angiography demonstrating a severely dilated left coronary artery to right atrial fistula (LCA-RA fistula). There are small branches of the LCA emerging from the fistula. (*B*) A wire rail is created from the femoral artery to the femoral vein allowing for advancement of a sheath from the right atrial into the fistula. (*C*) Placement of an Amplatzer duct occluder (ADO) into the fistula.

Ventricular Septal Defects

Although transcatheter device closure of ASD is widely accepted and now considered standard of care (discussed elsewhere in this article), closure of a VSD presents certain challenges that have required careful device modification. These challenges include the variable thickness of the ventricular septum, the variable location of many VSDs, the high pressures in the ventricles, and the close proximity of the aortic valve and conduction tissue to the membranous septum. VSD closure devices must avoid interference with valve function and the conduction system, and they must not produce arrhythmias, hemolysis, or device migration.[18]

The Amplatzer muscular VSD occluder (Abbott Laboratories, Chicago, IL) is in many ways similar to the Amplatzer ASD occlusion device; a Dacron polyester patch placed inside 2 nitinol disks connected by a waist. The Amplatzer muscular VSD occluder has demonstrated safety and efficacy in the closure of congenital and acquired (eg, after a myocardial infarction) muscular VSDs.[85–88] Other Amplatzer occlusion devices are also used for VSD closure, closure of ruptured sinus of Valsalva aneurysms (see **Fig. 6**) and other high-flow communications (**Fig. 7**), including the Amplatzer Duct Occluder, the vascular plugs, and the Amplatzer septal occluder. Closure of perimembranous VSDs is feasible but the development of conduction abnormalities is a major concern[89]

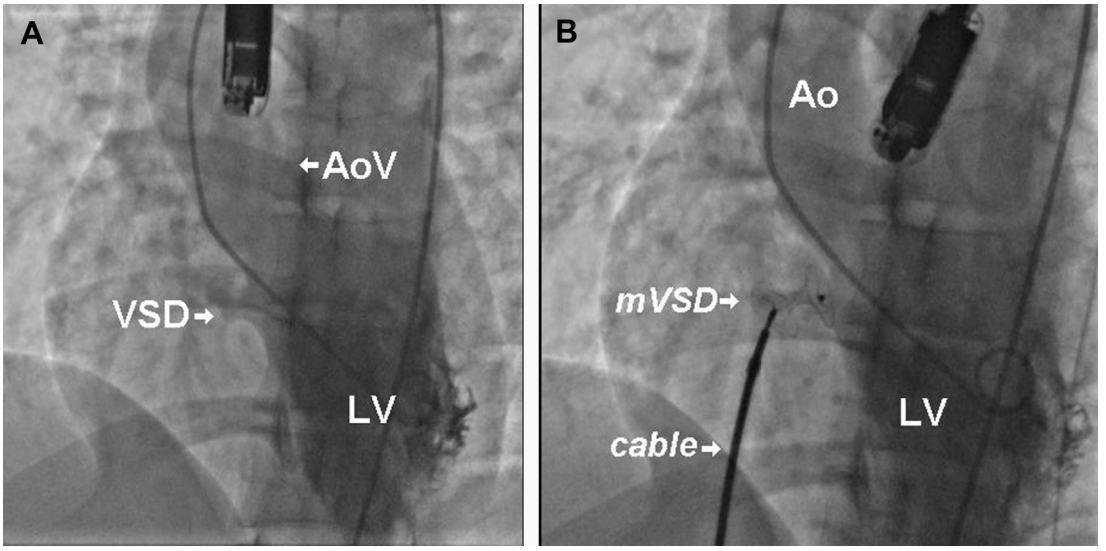

Fig. 9. (*A*) Left ventricular (LV) angiography demonstrating right to left shunting via a perimembranous VSD below the level of the aortic valve (AoV). (*B*) LVA angiography after muscular VSD device (mVSD) placement. The device is still attached to the delivery cable.

(Figs. 8 and 9). Closure of membranous VSDs associated with aneurysms of the ventricular septum is feasible and safe with a low risk of conduction system abnormalities.[90]

SUMMARY

Transcatheter interventions have become an indispensable tool in the treatment of adults with CHD. From closure of intracardiac and extracardiac communications to replacement of dysfunctional valves, the growing armamentarium of transcatheter tools is supplanting traditional surgical approaches. The next decade promises exponential advances in valve replacement and repair techniques, further refinement of stent and occlusion device technology, and an expanding role for transcatheter treatment of adults with CHD.

DISCLOSURE

Dr Aboulhosn is a consultant and proctor for Edwards Lifesciences, a consultant for Medtronic Inc and Abbott Pharmaceuticals. Dr Hijazi is a consultant for Occlutech and Numed, he is a speaker for Venus and Medtech.

REFERENCES

1. Moons P, Bovijn L, Budts W, et al. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. Circulation 2010;122:2264–72.
2. Gilboa SM, Salemi JL, Nembhard WN, et al. Mortality resulting from congenital heart disease among children and adults in the United States, 1999 to 2006. Circulation 2010;122:2254–63.
3. King TD, Thompson SL, Steiner C, et al. Secundum atrial septal defect. Nonoperative closure during cardiac catheterization. JAMA 1976;235:2506–9.
4. Rubio V, Limon-Lason R. Treatment of pulmonary valvular stenosis and tricuspid stenosis using a modified catheter. Arch Inst Cordiol Mexico 1952; 23:183–92.
5. Kan JS, White RI Jr, Mitchell SE, et al. Percutaneous balloon valvuloplasty: a new method for treating congenital pulmonary-valve stenosis. N Engl J Med 1982;307:540–2.
6. Stanger P, Cassidy SC, Girod DA, et al. Balloon pulmonary valvuloplasty: results of the valvuloplasty and angioplasty of congenital anomalies registry. Am J Cardiol 1990;65:775–83.
7. McCrindle BW. Independent predictors of long-term results after balloon pulmonary valvuloplasty. Valvuloplasty and Angioplasty of Congenital Anomalies (VACA) Registry investigators. Circulation 1994;89: 1751–9.
8. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association task force on clinical practice guidelines. J Am Coll Cardiol 2019;73:e81–192.
9. Fawzy ME, Galal O, Dunn B, et al. Regression of infundibular pulmonary stenosis after successful balloon pulmonary valvuloplasty in adults. Cathet Cardiovasc Diagn 1990;21:77–81.
10. Allen HD, Beekman RH 3rd, Garson A Jr, et al. Pediatric therapeutic cardiac catheterization: a statement for healthcare professionals from the Council on cardiovascular disease in the young, American Heart Association. Circulation 1998;97:609–25.
11. Hayes CJ, Gersony WM, Driscoll DJ, et al. Second natural history study of congenital heart defects. Results of treatment of patients with pulmonary valvar stenosis. Circulation 1993;87:I28–37.
12. Rao PS. Transcatheter treatment of pulmonary outflow tract obstruction: a review. Prog Cardiovasc Dis 1992;35:119–58.
13. Ring JC, Kulik TJ, Burke BA, et al. Morphologic changes induced by dilation of the pulmonary valve anulus with overlarge balloons in normal newborn lambs. Am J Cardiol 1985;55:210–4.
14. Butto F, Amplatz K, Bass JL. Geometry of the proximal pulmonary trunk during dilation with two balloons. Am J Cardiol 1986;58:380–1.
15. Sharieff S, Shah-e-Zaman K, Faruqui AM. Short- and intermediate-term follow-up results of percutaneous transluminal balloon valvuloplasty in adolescents and young adults with congenital pulmonary valve stenosis. J Invasive Cardiol 2003;15:484–7.
16. Rao PS, Galal O, Patnana M, et al. Results of three to 10 year follow up of balloon dilatation of the pulmonary valve. Heart 1998;80:591–5.
17. O'Connor BK, Beekman RH, Lindauer A, et al. Intermediate-term outcome after pulmonary balloon valvuloplasty: comparison with a matched surgical control group. J Am Coll Cardiol 1992;20:169–73.
18. Schneider DJ, Levi DS, Serwacki MJ, et al. Overview of interventional pediatric cardiology in 2004. Minerva Pediatr 2004;56:1–28.
19. Lock JE, Castaneda-Zuniga WR, Fuhrman BP, et al. Balloon dilation angioplasty of hypoplastic and stenotic pulmonary arteries. Circulation 1983;67:962–7.
20. Nakanishi T, Tobita K, Sasaki M, et al. Intravascular ultrasound imaging before and after balloon angioplasty for pulmonary artery stenosis. Catheter Cardiovasc Interv 1999;46:68–78.
21. Nakanishi T. Balloon dilatation and stent implantation for vascular stenosis. Pediatr Int 2001;43:548–52.
22. Magee AG, Wax D, Saiki Y, et al. Experimental branch pulmonary artery stenosis angioplasty using a novel cutting balloon. Can J Cardiol 1998;14: 1037–41.

23. Khairy P, Aboulhosn J, Gurvitz MZ, et al. Arrhythmia burden in adults with surgically repaired tetralogy of Fallot: a multi-institutional study. Circulation 2010; 122:868–75.
24. Lurz P, Nordmeyer J, Giardini A, et al. Early versus late functional outcome after successful percutaneous pulmonary valve implantation: are the acute effects of altered right ventricular loading all we can expect? J Am Coll Cardiol 2011;57:724–31.
25. Kaza AK, Lim HG, Dibardino DJ, et al. Long-term results of right ventricular outflow tract reconstruction in neonatal cardiac surgery: options and outcomes. J Thorac Cardiovasc Surg 2009;138:911–6.
26. Lluri G, Levi DS, Miller E, et al. Incidence and outcome of infective endocarditis following percutaneous versus surgical pulmonary valve replacement. Catheter Cardiovasc Interv 2018;91:277–84.
27. McElhinney DB, Benson LN, Eicken A, et al. Infective endocarditis after transcatheter pulmonary valve replacement using the Melody valve: combined results of 3 prospective North American and European studies. Circ Cardiovasc Interv 2013;6:292–300.
28. Sadeghi S, Wadia S, Lluri G, et al. Risk factors for infective endocarditis following transcatheter pulmonary valve replacement in patients with congenital heart disease. Catheter Cardiovasc Interv 2019;94: 625–35.
29. McElhinney DB, Sondergaard L, Armstrong AK, et al. Endocarditis after transcatheter pulmonary valve replacement. J Am Coll Cardiol 2018;72:2717–28.
30. Gillespie MJ, Rome JJ, Levi DS, et al. Melody valve implant within failed bioprosthetic valves in the pulmonary position: a multicenter experience. Circ Cardiovasc Interv 2012;5:862–70.
31. Finch W, Levi DS, Salem M, et al. Transcatheter melody valve placement in large diameter bioprostheses and conduits: what is the optimal "Landing zone"? Catheter Cardiovasc Interv 2015; 86:E217–23.
32. Shahanavaz S, Asnes JD, Grohmann J, et al. Intentional fracture of bioprosthetic valve frames in patients undergoing valve-in-valve transcatheter pulmonary valve replacement. Circ Cardiovasc Interv 2018;11:e006453.
33. Kenny D, Rhodes JF, Fleming GA, et al. 3-year outcomes of the Edwards SAPIEN transcatheter heart valve for conduit failure in the pulmonary position from the COMPASSION multicenter clinical trial. JACC Cardiovasc Interv 2018;11:1920–9.
34. Cheatham JP, Hellenbrand WE, Zahn EM, et al. Clinical and hemodynamic outcomes up to 7 years after transcatheter pulmonary valve replacement in the US melody valve investigational device exemption trial. Circulation 2015;131:1960–70.
35. Sinha S, Aboulhosn J, Asnes J, et al. Initial results from the off-label use of the SAPIEN S3 valve for percutaneous transcatheter pulmonary valve replacement: a multi-institutional experience. Catheter Cardiovasc Interv 2019;93:455–63.
36. McElhinney DB, Cheatham JP, Jones TK, et al. Stent fracture, valve dysfunction, and right ventricular outflow tract reintervention after transcatheter pulmonary valve implantation: patient-related and procedural risk factors in the US Melody Valve Trial. Circ Cardiovasc Interv 2011;4:602–14.
37. Ghobrial J, Levi DS, Aboulhosn J. Native right ventricular outflow tract transcatheter pulmonary valve replacement without pre-stenting. JACC Cardiovasc Interv 2018;11:e41–4.
38. Morgan GJ, Sadeghi S, Salem MM, et al. SAPIEN valve for percutaneous transcatheter pulmonary valve replacement without "pre-stenting": a multi-institutional experience. Catheter Cardiovasc Interv 2019;93:324–9.
39. Morray BH, McElhinney DB, Cheatham JP, et al. Risk of coronary artery compression among patients referred for transcatheter pulmonary valve implantation: a multicenter experience. Circ Cardiovasc Interv 2013;6:535–42.
40. Suleiman T, Kavinsky CJ, Skerritt C, et al. Recent development in pulmonary valve replacement after tetralogy of Fallot repair: the emergence of hybrid approaches. Front Surg 2015;2:22.
41. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: executive summary: a report of the American College of Cardiology/ American Heart Association Task Force on clinical practice guidelines. J Am Coll Cardiol 2019;73: 1494–563.
42. Singer MI, Rowen M, Dorsey TJ. Transluminal aortic balloon angioplasty for coarctation of the aorta in the newborn. Am Heart J 1982;103:131–2.
43. Forbes TJ, Kim DW, Du W, et al. Comparison of surgical, stent, and balloon angioplasty treatment of native coarctation of the aorta: an observational study by the CCISC (Congenital Cardiovascular Interventional Study Consortium). J Am Coll Cardiol 2011;58:2664–74.
44. Lock JE, Bass JL, Amplatz K, et al. Balloon dilation angioplasty of aortic coarctations in infants and children. Circulation 1983;68:109–16.
45. Kan JS, White RI Jr, Mitchell SE, et al. Treatment of restenosis of coarctation by percutaneous transluminal angioplasty. Circulation 1983;68:1087–94.
46. McCrindle BW, Jones TK, Morrow WR, et al. Acute results of balloon angioplasty of native coarctation versus recurrent aortic obstruction are equivalent. Valvuloplasty and Angioplasty of Congenital Anomalies (VACA) Registry Investigators. J Am Coll Cardiol 1996;28:1810–7.
47. Suarez de Lezo J, Pan M, Romero M, et al. Balloon-expandable stent repair of severe coarctation of aorta. Am Heart J 1995;129:1002–8.

48. Fletcher SE, Nihill MR, Grifka RG, et al. Balloon angioplasty of native coarctation of the aorta: midterm follow-up and prognostic factors. J Am Coll Cardiol 1995;25:730–4.
49. Ovaert C, McCrindle BW, Nykanen D, et al. Balloon angioplasty of native coarctation: clinical outcomes and predictors of success. J Am Coll Cardiol 2000; 35:988–96.
50. Taggart NW, Minahan M, Cabalka AK, et al. Immediate outcomes of covered stent placement for treatment or prevention of aortic wall injury associated with coarctation of the aorta (COAST II). JACC Cardiovasc Interv 2016;9:484–93.
51. Krichenko A, Benson LN, Burrows P, et al. Angiographic classification of the isolated, persistently patent ductus arteriosus and implications for percutaneous catheter occlusion. Am J Cardiol 1989;63: 877–80.
52. Fisher RG, Moodie DS, Sterba R, et al. Patent ductus arteriosus in adults–long-term follow-up: nonsurgical versus surgical treatment. J Am Coll Cardiol 1986;8: 280–4.
53. Pas D, Missault L, Hollanders G, et al. Persistent ductus arteriosus in the adult: clinical features and experience with percutaneous closure. Acta Cardiol 2002;57:275–8.
54. Roy A, Juneja R, Saxena A. Use of Amplatzer duct occluder to close severely hypertensive ducts: utility of transient balloon occlusion. Indian Heart J 2005; 57:332–6.
55. Balzer DT, Spray TL, McMullin D, et al. Endarteritis associated with a clinically silent patent ductus arteriosus. Am Heart J 1993;125:1192–3.
56. Parthenakis FI, Kanakaraki MK, Vardas PE. Images in cardiology: silent patent ductus arteriosus endarteritis. Heart 2000;84:619.
57. Schneider DJ, Moore JW. Patent ductus arteriosus. Circulation 2006;114:1873–82.
58. Moore JW, Khan M. Gianturco coil occlusion of patent ductus arteriosus. Curr Interv Cardiol Rep 2001; 3:80–5.
59. Pass RH, Hijazi Z, Hsu DT, et al. Multicenter USA Amplatzer patent ductus arteriosus occlusion device trial: initial and one-year results. J Am Coll Cardiol 2004;44:513–9.
60. Masura J, Tittel P, Gavora P, et al. Long-term outcome of transcatheter patent ductus arteriosus closure using Amplatzer duct occluders. Am Heart J 2006;151. 755.e7- 755.e10.
61. Eicken A, Balling G, Gildein HP, et al. Transcatheter closure of a non-restrictive patent ductus arteriosus with an Amplatzer muscular ventricular septal defect occluder. Int J Cardiol 2006;117(1):e40–2.
62. Spies C, Ujivari F, Schrader R. Transcatheter closure of a 22 mm patent ductus arteriosus with an Amplatzer atrial septal occluder. Catheter Cardiovasc Interv 2005;64:352–5.
63. Sadiq M, Malick NH, Qureshi SA. Simultaneous treatment of native coarctation of the aorta combined with patent ductus arteriosus using a covered stent. Catheter Cardiovasc Interv 2003;59:387–90.
64. Du ZD, Hijazi ZM, Kleinman CS, et al. Comparison between transcatheter and surgical closure of secundum atrial septal defect in children and adults: results of a multicenter nonrandomized trial. J Am Coll Cardiol 2002;39:1836–44.
65. Varma C, Benson LN, Silversides C, et al. Outcomes and alternative techniques for device closure of the large secundum atrial septal defect. Catheter Cardiovasc Interv 2004;61:131–9.
66. Cooke JC, Gelman JS, Harper RW. Echocardiologists' role in the deployment of the Amplatzer atrial septal occluder device in adults. J Am Soc Echocardiogr 2001;14:588–94.
67. Magni G, Hijazi ZM, Pandian NG, et al. Two- and three-dimensional transesophageal echocardiography in patient selection and assessment of atrial septal defect closure by the new DAS-Angel Wings device: initial clinical experience. Circulation 1997; 96:1722–8.
68. Carcagni A, Presbitero P. Transcatheter closure of secundum atrial septal defects with the Amplatzer occluder in adult patients. Ital Heart J 2002;3:182–7.
69. Butera G, Chessa M, Bossone E, et al. Transcatheter closure of atrial septal defect under combined transesophageal and intracardiac echocardiography. Echocardiography 2003;20:389–90.
70. Zanchetta M, Rigatelli G, Pedon L, et al. Transcatheter atrial septal defect closure assisted by intracardiac echocardiography: 3-year follow-up. J Interv Cardiol 2004;17:95–8.
71. Perloff JK. Ostium secundum atrial septal defect–survival for 87 and 94 years. Am J Cardiol 1984; 53:388–9.
72. Campbell M. Natural history of atrial septal defect. Br Heart J 1970;32:820–6.
73. Abdelkarim A, Levi DS, Tran B, et al. Fenestrated Transcatheter ASD closure in adults with diastolic dysfunction and/or pulmonary hypertension: case series and review of the literature. Congenit Heart Dis 2016;11:663–71.
74. Schwerzmann M, Zafar M, McLaughlin PR, et al. Atrial septal defect closure in a patient with "irreversible" pulmonary hypertensive arteriopathy. Int J Cardiol 2006;110:104–7.
75. Bradley EA, Ammash N, Martinez SC, et al. Treat-to-close": non-repairable ASD-PAH in the adult: results from the North American ASD-PAH (NAAP) multicenter registry. Int J Cardiol 2019;291:127–33.
76. Banerjee A, Bengur AR, Li JS, et al. Echocardiographic characteristics of successful deployment of the Das AngelWings atrial septal defect closure device: initial multicenter experience in the United States. Am J Cardiol 1999;83:1236–41.

77. Walsh KP, Tofeig M, Kitchiner DJ, et al. Comparison of the Sideris and Amplatzer septal occlusion devices. Am J Cardiol 1999;83:933–6.
78. Du ZD, Koenig P, Cao QL, et al. Comparison of transcatheter closure of secundum atrial septal defect using the Amplatzer septal occluder associated with deficient versus sufficient rims. Am J Cardiol 2002;90:865–9.
79. Rao PS. Comparative summary of atrial septal defect occlusion devices. Philadelphia: Lippincott, Williams & Wilkins; 2003.
80. 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;45:505–7.
81. Lopez K, Dalvi BV, Balzer D, et al. Transcatheter closure of large secundum atrial septal defects using the 40 mm Amplatzer septal occluder: results of an international registry. Catheter Cardiovasc Interv 2005;66:580–4.
82. de Hemptinne Q, Horlick EM, Osten MD, et al. Initial clinical experience with the GORE((R)) CARDIOFORM ASD occluder for transcatheter atrial septal defect closure. Catheter Cardiovasc Interv 2017;90:495–503.
83. Kumar P, Orford JL, Tobis JM. Two cases of pericardial tamponade due to nitinol wire fracture of a gore septal occluder. Catheter Cardiovasc Interv 2019. https://doi.org/10.1002/ccd.28596.
84. Amin Z, Hijazi ZM, Bass JL, et al. Erosion of Amplatzer septal occluder device after closure of secundum atrial septal defects: review of registry of complications and recommendations to minimize future risk. Catheter Cardiovasc Interv 2004;63:496–502.
85. Chessa M, Carminati M, Cao QL, et al. Transcatheter closure of congenital and acquired muscular ventricular septal defects using the Amplatzer device. J Invasive Cardiol 2002;14:322–7.
86. Thanopoulos BD, Karanassios E, Tsaousis G, et al. Catheter closure of congenital/acquired muscular VSDs and perimembranous VSDs using the Amplatzer devices. J Interv Cardiol 2003;16:399–407.
87. Holzer R, Balzer D, Cao QL, et al. Device closure of muscular ventricular septal defects using the Amplatzer muscular ventricular septal defect occluder: immediate and mid-term results of a U.S. registry. J Am Coll Cardiol 2004;43:1257–63.
88. Holzer R, Balzer D, Amin Z, et al. Transcatheter closure of postinfarction ventricular septal defects using the new Amplatzer muscular VSD occluder: results of a U.S. Registry. Catheter Cardiovasc Interv 2004;61:196–201.
89. Holzer R, de Giovanni J, Walsh KP, et al. Transcatheter closure of perimembranous ventricular septal defects using the Amplatzer membranous VSD occluder: immediate and midterm results of an international registry. Catheter Cardiovasc Interv 2006;68:620–8.
90. Pedra CA, Pedra SR, Esteves CA, et al. Percutaneous closure of perimembranous ventricular septal defects with the Amplatzer device: technical and morphological considerations. Catheter Cardiovasc Interv 2004;61:403–10.