Cardiology in the Young (2017), **27**, 1974–1985
doi:10.1017/S1047951117002141

Original Article

Landmark lecture on interventional cardiology: interventional cardiac catheterisation for CHD: the past, present, and the future*

Ziyad M. Hijazi,[1,2] Damien Kenny[3]

[1]*Sidra Cardiac Program, Sidra Medical & Research Center, Doha, Qatar;* [2]*Weill Cornell Medicine, New York, New York, United States of America;* [3]*Our Lady's Children's Hospital, Dublin, Ireland*

Abstract CHD affects millions of patients worldwide. Interventional therapies for CHD goes back to the mid-1960s when Bill Rashkind performed balloon atrial septostomy on a cyanotic baby with transposition of the great vessels. This was followed by development of balloon catheters to perform balloon valvuloplasties and angioplasties in the early to late 1980s. Although King and Mills performed the first transcatheter closure of secundum atrial septal defect in the mid-1970s, this procedure was better realised in the mid-1990s. More intracardiac defect closures were performed in the late 1990s and early 2000. This brings us to the current era of percutaneous valve implantation as developed by Bonhoeffer. In this paper, we will discuss the past, present, and future of interventional cardiac catheterisation for CHD patients.

Keywords: Cardiac catheterization; congenital heart disease; transcatheter pulmonary valve replacement

Received: 15 September 2017; Accepted: 5 December 2017

From the time Rashkind performed the first interventional procedure, balloon atrial septostomy in CHD to palliate an infant with transposition of the great vessels, interventional therapies to treat or palliate infants, children, and adults with CHD have come a long way. In this article, I will discuss the milestones of this discipline highlighting the procedures we have been performing and continue to perform currently. I will divide the era of interventional therapies into three distinct eras: the past – balloon atrial septostomy, percutaneous balloon valvuloplasty, and balloon angioplasty/stent implantation for vessel narrowing – the present – septal defect closure under two-dimensional and three-dimensional echocardiographic guidance, hybrid interventions for various conditions – and the future of cardiac catheterisation – percutaneous pulmonary valve implantation, bioresorbable stent implantation and Fontan completion in the catheterisation laboratory.

The past

Balloon atrial septostomy

Balloon atrial septostomy first described by Rashkind[1] is needed when there is restricted communication between the right and left atria. A few techniques have been developed to create the interatrial communication. These include balloon atrial septostomy; blade atrial septostomy; static balloon dilation of the septum; and radiofrequency perforation or transseptal puncture to create the communication and then enlarge it using one of the above techniques.

Balloon atrial septostomy can be performed in the cardiac catheterisation laboratory or bed-side under echocardiographic guidance. Possible complications could include the following: transient rhythm disturbances, which may be permanent or even fatal; failure to create adequate atrial communication – due to thick septum; balloon fragment embolisation; laceration of the atrioventricular valves; and laceration of systemic or pulmonary veins.

*Presented at the 2017 Seventh World Congress of Pediatric Cardiology & Cardiac Surgery (WCPCCS 2017), Barcelona, Spain, 16–21 July, 2017. Presented Friday, 21 July, 2017.

Correspondence to: Z. M. Hijazi, MD, MPH, Chair, Department of Pediatrics, Sidra Cardiac Program, Sidra Medical & Research Center, Doha, Qatar. Tel: +974 4003 6602; E-mail: zhijazi@sidra.org

Blade atrial septostomy and static balloon dilation. In neonates with thick or intact atrial septum, blade septostomy is needed before balloon septostomy. However, currently, most centres rely on static balloon – regular balloon or cutting balloon – dilation to enlarge the communication as it is very effective when a controlled diameter of the opening is desired.

Atrial septal stenting. Atrial septal stenting is indicated in cases of thick septum and is subject to recoil. Furthermore, if the opening is required for a longer period of time, stenting the defect may offer this benefit.

Balloon valvuloplasty

Pulmonary valvuloplasty. First performed by Kan et al,[2] balloon pulmonary valvuloplasty remains the treatment of choice for valvar pulmonary stenosis at all ages. Critical pulmonary stenosis may be lethal in neonates. Indications for pulmonary valvuloplasty include symptomatic patient, asymptomatic patient with systolic gradient $\geqslant$40 mmHg across the pulmonary valve, right ventricular hypertrophy and dysfunction, as a palliative procedure in a patient with complex cyanotic heart disease including some rare cases of Tetralogy of Fallot.[3,4] The main complication of the procedure is pulmonary regurgitation, which may present at a later stage during a longer-term follow-up. To decrease the risk for pulmonary regurgitation, it is better to use smaller balloon to annulus ratio than the previously recommended 120–140%. In selected cases with pulmonary atresia, multiple techniques have been used to perforate the valve, such as using the stiff end of the guide wire, laser, and most commonly, radiofrequency wires. Complications encountered include perforation of the right ventricular outflow tract leading to pericardial effusion and tamponade. One group that clearly needs to be excluded from this procedure are patients with right ventricular-dependent coronary circulation.

Aortic valvuloplasty. Balloon aortic valvuloplasty first described by Lababidi[5] has become a safe and effective treatment of valvar aortic stenosis. It is indicated in the newborn with isolated critical valvar stenosis defined as ductal dependent circulation with congenital aortic stenosis.[3] Moreover, it is indicated in isolated valvar aortic stenosis with depressed LV function, with congestive heart failure or shock, or with a peak to peak gradient of $\geqslant$50 mmHg.[3] The initial balloon chosen should be 85–90% of the aortic annulus measured by angiography. Optimal result should achieve at least 50% reduction in the gradient with no increase in aortic regurgitation or residual gradient of less than 35 mmHg and absence of regurgitation. Complications of the procedure include suboptimal relief of the obstruction, creation of moderate or severe aortic incompetence, femoral artery injury, thrombo-embolic stroke, injury to mitral valve, and perforation of the myocardium.

Mitral balloon valvuloplasty. Congenital mitral stenosis encompasses a broad spectrum of anatomic variants, including the typical variants with thickened leaflets, shortened chordae and decreased interchordal spaces; supra-mitral valve ring; and double mitral orifices, parachute mitral valve and the hypoplastic mitral valve associated with hypoplastic left heart syndrome. Balloon valvuloplasty is rarely performed for MS. Torn leaflets and chordal attachments and papillary muscle rupture are the most common cause of regurgitation after dilation.

Balloon angioplasty and/or stent placement

Native coarctation and recoarctation. Coarctation accounts for ~6–8% of all cardiac defects. The usual location of caorctation is juxtaductal, just distal to the left subclavian artery. Neonates may present with signs and symptoms of low cardiac output and shock as the ductus arteriosus closes. A pressure gradient between upper and lower limbs and weak femoral pulse are suggestive of coarctation.

In recent years, balloon angioplasty has emerged as an alternative, less invasive option to the gold standard surgical therapy in infants with discrete narrowing and no evidence of arch hypoplasia and in cases of recoarctation after surgery or dilatation. The complications of the procedure are mainly recurrence of stenosis and a small but important incidence of aortic aneurysm. Stent implantation for native coarctation or recoarctation of the aorta has also emerged as a beneficial therapeutic option for patients who can receive a stent that can be expanded to an adult size.[3]

Pulmonary artery stenosis. The overall frequency of peripheral arterial stenosis is 2–3%. It is either an isolated lesion or associated with other defects or part of some syndromes.

Severe pulmonary arterial stenosis is obvious when a pressure gradient of >20–30 mmHg is found or elevation of right ventricular or proximal main pulmonary artery pressure is greater than one-half to two-thirds of systemic pressure secondary to the more distal obstruction or when there is relative flow discrepancy between the two lungs of 35%/65% or worse.[3,4]

Treatment of pulmonary artery stenosis depends on the site of the stenotic segments. Percutaneous pulmonary angioplasty is suitable for distal lesions unreachable by surgery. Different techniques of percutaneous treatment include balloon angioplasty,

cutting balloons angioplasty and intravascular stent placement.[4] The major risk for pulmonary angioplasty includes vessel perforation.

Systemic and pulmonary veins balloon angioplasty. Systemic venous stenosis occurs as a congenital defect or subsequent to surgery or from indwelling lines in the veins or from external compression. No significant gradient could be detected because of low-pressure system. Surgery of systemic venous stenosis is difficult and unrewarding. Balloon dilation, with or without stent implantation, had proved effective with little morbidity and mortality. Pulmonary vein stenosis can be a congenital lesion or acquired after corrective surgery or after lung transplantation. The treatment modalities include surgery, balloon angioplasty, use of cutting balloon and, as last resort, stenting in older babies. Restenosis rates after angioplasty and stent are high and particularly worse for congenital pulmonary vein stenosis. Potential complications include vessel dissection, stent malposition and embolisation. There is a high rate of restenosis that can make the procedure almost futile.[6]

The present

Septal defect closure

Patent ductus arteriosus closure. This represents the second oldest interventional procedure in CHD, when Porstmann et al performed closure of a patent ductus arteriosus in 1966 and reported on it in 1967.[7] Occlusion of patent ductus arteriosus is indicated when neonates are symptomatic due to circulatory overload. Compared with the time when Porstmann reported the successful trial of patent ductus arteriosus occlusion, today a variety of devices and innovative techniques are available for transcatheter occlusion of patent ductus arteriosus. Coils are used for patent ductus arteriosus occlusion and their use in small patent ductus arteriosus of <2 mm in diameter has become the treatment of choice. In larger ducts, many use devices – Amplatzer duct occlude among others – to close such patent ductus arteriosus. Potential risks include inadvertent device embolisation into the pulmonary or systemic artery, device obstruction to aortic or pulmonary flow, especially in small infants, transient left ventricular systolic dysfunction, and haemolysis and recanalisation.

Atrial septal defect closure. Atrial septal defects are among the most common CHD and they account for one-third of CHD in adults. Significant iatrogenic atrial septal defects also have been described after adopting new modalities of percutaneous transcatheter therapies to the left side of the heart (e.g. mitral clip for mitral insufficiency). At present, only the secundum type is amenable for transcatheter closure. Most children with an atrial septal defect present with a murmur and are asymptomatic. Occasionally, infants may present with breathlessness, recurrent chest infections, and even heart failure. Failure to thrive is an uncommon presentation.

The clinical course of an unrepaired atrial septal defect in adulthood may be significantly affected by hypertension, coronary artery disease, and mitral regurgitation. Patients with unrepaired atrial septal defect over 60 years of age often develop atrial fibrillation, an age-related reflection of atrial stretch, which seldom occurs in those younger than 40 years of age.[8] In the current era of percutaneous device closure of interatrial communications, evaluation with transesophageal echocardiography specially if accompanied with real-time three-dimensional imaging or intracardiac echocardiography is mandatory before considering device closure.[9]

Initial screening imaging with transthoracic echocardiography may demonstrate a clearly visible defect in the atrial septum, best seen in the apical four chamber and subcostal long-axis views. However, it is common to see "echo dropout" in the region of the interatrial septum, and this may lead to misdiagnosis. Cardiac catheterisation is not usually required for diagnosis. However, in older patients a diagnostic cardiac catheterisation for coronary angiography and haemodynamic assessment may be justified. Patients with high left ventricular end-diastolic pressure (>14 mmHg) need medical optimisation before closure.[10] The most widely used device to close the atrial septal defect is the Amplatzer septal occluder. There are other double-desk devices available to close such defects, which include the following: Occlutech Figulla Flex-II device, Gore Cardioform Septal occluder, and a variety of Amplatzer knock out devices.

Closure of secundum atrial septal defect is now considered the treatment of choice for eligible defects and it has been proven to be safer than surgery.[11] However, there are potential complications that the operator and cardiologist need to be aware of. These include the following: device embolisation (1–2%), thromboembolic episodes – transient ischaemic attack, stroke – [<1%], arrhythmias, heart block – this is a rare occurrence, air embolism, erosion – rare at about 2–3/1000 cases, pericardial effusion, death – extremely rare at about 2/10,000, and endocarditis – very rare.

Ventricular septal defect closure. Ventricular septal defect represents the most common CHD, accounting for ~40% of all cardiac defects. Ventricular septal defect can be located in the muscular septum or in the membranous septum.

https://doi.org/10.1017/S1047951117002141 Published online by Cambridge University Press

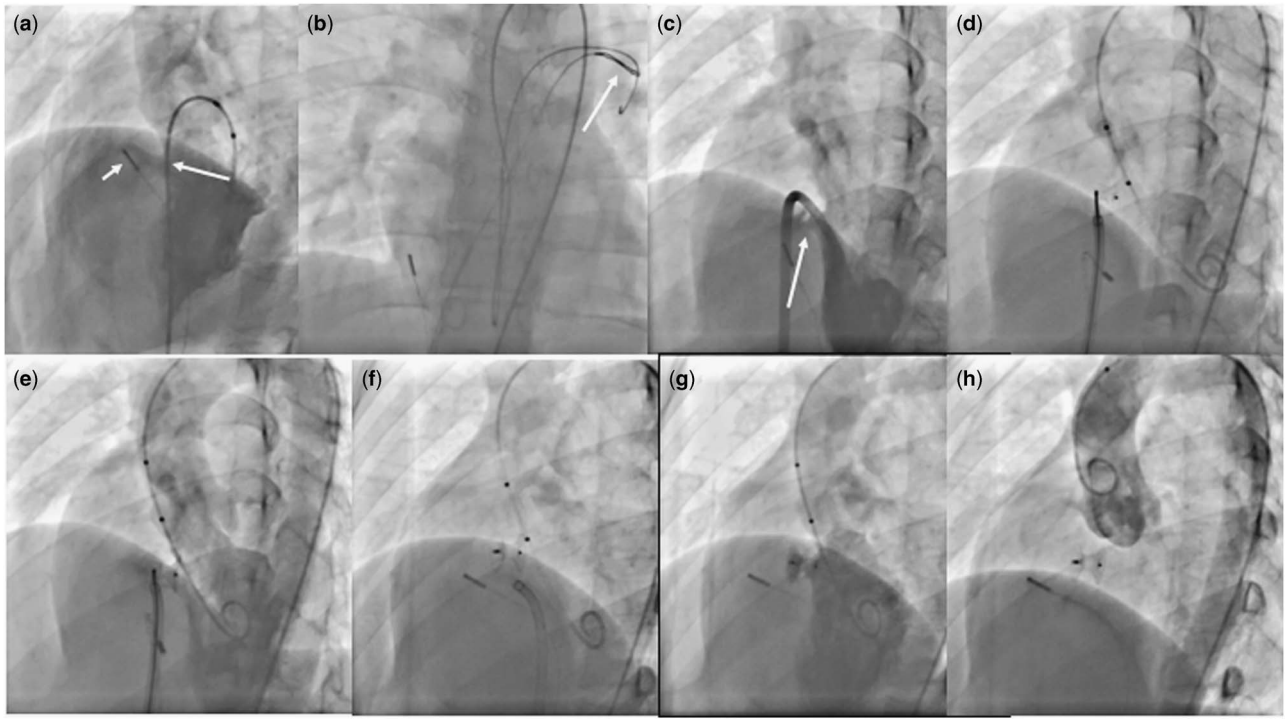

Figure 1.
*Transcatheter perimembranous ventricular septal defect closure using a muscular ventricular septal defect device. (**a**) Angiogram in left anterior oblique view (55° left anterior oblique/20° cranial) showing a moderate-sized membranous ventricular septal defect (long arrow) with good distance from the aortic valve. Note the presence of intracardiac echo catheter (short arrow). (**b**) Cine fluoroscopy during snaring of a guide wire (arrow) (from the left ventricle through the ventricular septal defect to left pulmonary artery in the left pulmonary artery). (**c**) A 8 Fr delivery sheath is positioned from the femoral vein to right atrium, right ventricle via defect into the left ventricle. (**d**) A 8 mm Amplatzer muscular ventricular septal defect occluder is deployed but not released across the defect. (**e**) Angiogram in left ventricle documenting good device position before release. (**f**) Device has just been released. (**g**) Left ventricle angiogram demonstrating good device position and minimal foaming through the device. (**h**) Ascending aorta angiogram showing good distance from the edge of device.*

Closure of muscular ventricular septal defect is an approved procedure using the Amplatzer muscular ventricular septal defect device. Transcatheter closure provides safe and effective therapy specially for multiple ventricular septal defects – Swiss cheese defects.[12,13] Closure should be carried out under fluoroscopic and echocardiographic – transthoracic, transesophageal, or intracardiac – guidance. Closure of perimembranous ventricular septal defect can be done; however, the complication rates have been unacceptably higher compared with surgical closure.[14] Therefore, currently, only selected cases can be done with acceptable results. The higher incidence of complete heart block (5.5%) and new aortic valve insufficiency using the Amplatzer membranous ventricular septal defect occluder pushed the investigators and manufacturer of the device to terminate the trial, hoping for a new design of the device to provide the answer. The new design is still awaiting confirmation in a clinical trial. Figure 1 demonstrates closure of a moderate-sized perimembranous ventricular septal defect in a 23-year-old young patient that was closed using an 8 mm Amplatzer Muscular ventricular septal defect device.

Hybrid interventions

Hybrid closure of muscular ventricular septal defect. Studies have documented a higher incidence of adverse events as well as lower procedural success when ventricular septal defect closure is performed percutaneously in young patients with a weight below 5–10 kg.[12,15] The difficulties in small children relate to the establishment of an arteriovenous wire loop, which results in splinting of the tricuspid and/or aortic valves, which most likely will result in haemodynamic disturbance, which is poorly tolerated. In addition, the limited space can lead to kinks within the delivery sheath that make advancing the device cumbersome and difficult. Furthermore, haemodynamic instability can be further aggravated through blood loss during the procedure, which for a young patient can be significant. For all the above reasons, percutaneous closure is usually not recommended in young

https://doi.org/10.1017/S1047951117002141 Published online by Cambridge University Press

patients, and instead a perventricular "Hybrid" approach should be considered, which has been successfully used with low morbidity and mortality.[16] It is an approach that has been increasingly utilised in many institutions, with the C3PO registry documenting a significant increase in Hybrid procedures being performed.[17] In this context, hybrid perventricular ventricular septal defect closure is one of the most common hybrid procedures, and carried out through cooperation between cardiothoracic surgeons and interventional cardiologists using either a dedicated hybrid catheterisation laboratory or a cardiac surgical operating room. The procedure is carried out on the beating heart without the use of cardiopulmonary bypass. The procedure can be carried out primarily under echocardiographic guidance – transesophageal echocardiography or epicardial, or in the presence of multiple muscular ventricular septal defects, it can be done under both echocardiographic and fluoroscopic guidance. Using a sub-xiphoid incision an appropriate entry site is identified under echo guidance at the right ventricular free wall, ideally perpendicular to the ventricular septal defect, but allowing a sufficient distance to deploy the device. Once a purse string suture has been placed, a direct puncture is performed with the needle directed toward the ventricular septal defect. Most frequently, an 0.035" angled glide-wire is directed under echo guidance across the ventricular septal defect. Once wire position is confirmed within the left ventricular cavity, an appropriately sized short sheath is advanced over the wire into the left ventricle. It is important to retract the dilator a little and be conscious of the distance to the left ventricular posterior free wall. Advancing the sheath/dilator too far can create an injury at the left ventricular wall, either directly with the sheath/dilator, or when advancing/deploying the device and must be avoided. Echo guidance is crucial in this process. Patients with associated cardiac defects – coarctation of the aorta, transposition, etc. – requiring treatment can undergo repair after the ventricular septal defect(s) is closed.

Hybrid management of hypoplastic left heart syndrome. Despite several innovative approaches designed to address the major suspected culprits leading to mortality, survival is consistently reported at between 65% and 70% at 5 years.[18,19] Even a modern-day cohort of neonates with hypoplastic left heart syndrome randomised into Norwood or Sano surgical palliation carries a heavy mortality burden at 2 years.[20] Within the cohort of survivors, concerns have been raised regarding longer-term neurodevelopmental outcomes secondary to prolonged cardiopulmonary bypass with circulatory arrest in the neonatal period.[20] It is within this historical context that the hybrid approach to the treatment of hypoplastic left heart syndrome evolved. The main caveats to this approach were to maintain adequate perfusion pressure to the systemic circulation across the arterial duct following the natural reduction in pulmonary vascular resistance after birth. The precursors to achieving this were supported by initial technical experience with stenting the arterial duct to maintain pulmonary circulation in the setting of pulmonary atresia.[21,22] Subsequently, this approach was employed in the setting of hypoplastic left heart syndrome as a bridge to transplantation.[23] Although this strategy achieved its short-term goal, exclusive stenting of the arterial duct would eventually lead to pulmonary overflow and "steal" from the systemic circulation and thus further modifications were necessary for longer-term palliation. The first complete hybrid palliative approach to hypoplastic left heart syndrome was described in 1993 by Gibbs et al, with surgical banding of the branch pulmonary arteries and percutaneous stenting of the arterial duct.[24] This was coupled with either surgical or percutaneous decompression of the left atrium by opening the atrial septum. Thus, the three main physiological objectives of hypoplastic left heart syndrome palliation, namely unobstructed systemic output, balanced pulmonary circulation, and an unrestrictive atrial communication, were addressed. Although there was no procedural mortality, two of the four patients died within 2 weeks because of excessive pulmonary blood flow and subsequent right ventricular failure, with prolonged hospitalisation of the third patient due to similar concerns. Subsequent reports of extended experience with longer-term follow-up from the same group confirmed these initial concerns with no survivors beyond 30 months from all eight patients palliated with this approach.[25] The cited reason for demise in five of the eight patients was pulmonary over circulation with subsequent right ventricular failure. It was not clear from this report whether there were consistent targets for determining effective pulmonary artery banding; however, it was clear that pulmonary over circulation was not tolerated in this group. Further experience with the hybrid approach was not described until 2002 when the group from Giessen reported clearer pre-procedural targets for the tightness of the pulmonary artery band with the goal to achieve distal pulmonary artery pressures <50% of systemic pressures, confirmed by Doppler velocities of greater than 4 m/second across the bands and reduction in systemic oxygen saturations to ~80%.[26] Ten of the 11 patients survived to a stage 2 procedure, which was heart transplantation in two and a modified Norwood procedure in the remaining eight patients. There was one intraoperative death with overall survival through stage 2 of 82%. A further report from this group with experience in 58 patients confirmed these survival

https://doi.org/10.1017/S1047951117002141 Published online by Cambridge University Press

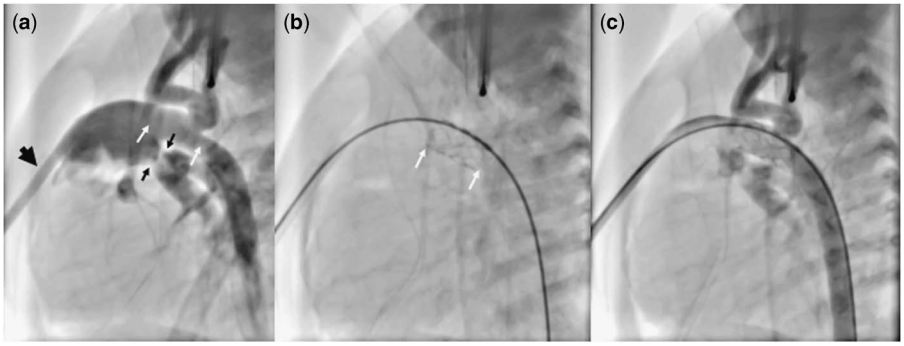

Figure 2.
*Hybrid pulmonary artery banding and patent ductus arteriosus stent in a neonate with hypoplastic left heart syndrome. (**a**) Angiogram via a sheath (black arrow head) inserted into the main pulmonary artery just above the pulmonic valve showing good bands across the left and right pulmonary arteries (black arrows: the top arrow is the left pulmonary artery; bottom one is the right pulmonary artery) and the large patent ductus arteriosus measuring ~5.8 mm. The boundary of the patent ductus arteriosus is marked by the two white arrows. (**b**) Cine fluoroscopy after deployment of a 8 × 15 mm stent (Genesis). The two white arrows denote the stent. (**c**) Final angiogram showing good stent position and the branch pulmonary arteries.*

figures.[27] Interestingly, the initial approach adopted by this group consisted of percutaneous ductal stenting followed by surgical pulmonary artery banding within 72 hours; however, stent dislodgement occurred in two patients because of manipulation at the time of surgery, and this approach was reversed so as to perform the pulmonary artery banding in the operating room followed by ductal stenting with either balloon expandable or self-expanding stents depending on the duct morphology. During this same time period, other investigators (the Columbus group) were attempting to achieve pulmonary artery flow restriction and unobstructed flow to the systemic circulation exclusively via a transcatheter approach.[28] This involved deployment of custom-designed nitinol flow restrictors in the pulmonary arteries; however, delivery was associated with significant haemodynamic instability and this approach was abandoned. This group further evolved their practice to describe the first truly hybrid approach to hypoplastic left heart syndrome with surgeon and interventionalist working side-by-side to band the pulmonary arteries and stent the arterial duct through the pulmonary artery with further transcatheter intervention to the atrial septum performed as a separate procedure if necessary. This group subsequently published their extended experience of 40 neonates with this approach[29] with 83% survival. This included 15 patients through stage 3 palliation. The hybrid approach to hypoplastic left heart syndrome has evolved into a distinct management strategy since then, with published reports on how to approach atrial septal interventions,[30] the retrograde aortic arch,[31] and anaesthetic,[32] and enteral feeding[33] management of these patients. Controversy still exists whether this approach should be offered as an alternative to Norwood surgery or limited to neonates where surgery has proved to be a substantially higher risk. Only randomised trials will answer this debate for once and all. Figure 2 depicts an angiogram of a 28-day young neonate with hypoplastic left heart syndrome who underwent a hybrid intervention – surgical band placement on the branch pulmonary artery and patent ductus arteriosus stent placement.

Intraoperative stent implantation. Indications for intraoperative stent implantation include the following: a need for a concomitant surgical procedure, limited vascular access, small patient size, and as rescue therapy after failed percutaneous stent placement. One important advantage of intraoperative or hybrid stent implantation is that regardless of patient size a stent ultimately capable of achieving an adult diameter can nearly always be implanted. The majority of children requiring stent implantation at any age would ideally receive stents capable of being expanded to an adult size. Techniques employed to deliver the stent can include: *videoscopic guided stent implantation*, which has been used solely for the treatment of branch pulmonary artery stenosis, and typically is a planned procedure that takes place in the operating room. The second technique, *stent implantation via surgically provided vascular access*, has been used to treat a wide variety of lesions including branch pulmonary artery stenoses, recurrent aortic arch obstruction – after hypoplastic left heart syndrome palliative surgery, and shunt occlusion/stenosis. This approach can be performed in either the catheterisation laboratory or surgical suite or if available a hybrid suite.[34]

Transcatheter pulmonary valve replacement

Since Bonhoeffer[35] performed the first human case of transcatheter pulmonary valve replacement, two

https://doi.org/10.1017/S1047951117002141 Published online by Cambridge University Press

valves have already been commercially approved for this purpose. Transcatheter pulmonary valve replacement is one of the most exciting recent developments in the treatment of CHD and has evolved as an attractive alternative to surgery in patients with dysfunctional right ventricle-pulmonary artery conduits or bioprosthetic valves. The most common indication of pulmonary valve implantation is residual right ventricular outflow tract lesion after repair of CHD that can be stenotic, regurgitant, or mixed. Tetralogy of Fallot remains the most common diagnosis for patients undergoing this procedure, followed by patients who underwent the Ross operation for aortic valve disease. Currently, there are two commercially available valves: the Melody (Medtronic Inc, Minneapolis, Minnesota, United States of America) and Sapien XT (Edwards Lifesciences, Irvine, California, United States of America) valves. Even though transcatheter pulmonary valve replacement is not currently a standard indication in patients with native right ventricular outflow tract [see section on future of cardiac catheterisation], many centres perform transcatheter pulmonary valve replacement in a select group of patients who have a native outflow tract – post-transannular patch repair of tetralogy of Fallot – that is of an appropriate size or one that can be altered to a suitable size by insertion of multiple stents.[36]

Appropriate patient selection for pulmonary valve replacement is crucial, and the guidelines for both surgical as well as transcatheter pulmonary valve replacement have continued to evolve over the last decade.[37–41] In general, it is recommended to use a composite measure of clinical data – symptoms, exercise capacity, presence of arrhythmias, – electrocardiogram and echo data, and MRI-based data elements such as right ventricular end diastolic volume index ($\geqslant$150 ml/m^2), right ventricular end-systolic volume index (>80 ml/m^2), pulmonary regurgitant fraction ($\geqslant$40%), and right ventricular function (ejection fraction <40%).[42,43] Pulmonary valve replacement should ideally be performed before right ventricular function declines. In addition to clinical indications, several anatomical criteria need to be fulfilled to qualify for transcatheter pulmonary valve replacement. The ideal anatomy for transcatheter pulmonary valve replacement is a uniform diameter from right ventricular outflow tract to pulmonary artery bifurcation – conduit between right ventricle and main pulmonary artery or a patient with a bioprostehtic valve between the right ventricle and pulmonary artery – with adequate main pulmonary artery length to avoid stenting into the pulmonary artery bifurcation. With the current iterations of the Melody valve, the right ventricular outflow tract, pulmonary valve annulus, and proximal main pulmonary artery must be 22 mm or less to prevent leaflet malcoaptation. Using the 22 mm Ensemble delivery system, the outer diameter of the Melody valve is ~24 mm, and therefore any inner diameter of a conduit larger than this would be insufficient to securely anchor the valve. Nevertheless, there is limited experience with mounting the Melody valve on a 24 mm balloon delivered through a 24-Fr sheath.[44] On the other hand, the SAPIEN XT valve can be deployed in right ventricular outflow tract sizes up to 29 mm in diameter. During cardiac catheterisation, routine haemodynamic data are obtained. Gradients are obtained in the right ventricular outflow tract/conduit, at the level of the pulmonary valve, and the pulmonary arteries. The pulmonary artery and branch morphology should be delineated by pulmonary arteriograms to define the architecture. After haemodynamic assessment, evaluation of the coronary arteries must be made with balloon inflation in the right ventricle outflow tract to assess the proximity of the coronary arteries to the outflow tract. When the conduit is placed on the anterior surface of the heart, coronary branches may pass directly beneath it, and may be potentially compressed by placement of the stented valve and distension of the conduit.[45] If no evidence of coronary compression is noted, pre-stenting of the right ventricular outflow tract using bare/covered metal stents is performed to create an appropriate landing zone for the transcatheter valve. Pre-stenting the landing site has significantly improved the survival of the implant, minimising stent fracture, which affected 23% of the initially reported cases of the Melody valve. The appropriate valve is then introduced and deployed. Figure 3 depicts a patient who received an Edwards valve for severe conduit dysfunction.

Serious complications associated with transcatheter pulmonary valve replacement are very rare, but are devastating when they happen. In the US multicenter SAPIEN study (COMPASSION), the rate of serious complications was as high as 19.4% in the initial 36 procedures attempted,[46] with no mortality encountered. However, in COMPASSION trial, with improved operator experience, after July, 2009 until 2014 – end of the trial, no further cases of valve migration had been reported. The Italian multicenter registry reported a major complication rate of 11%.[47] We expect that the complication rates will decrease significantly as operators become more experienced with the use of these valves. Expected complications can be broken down into several categories, including vascular complications, coronary compression, stent fracture,[48] homograft rupture, valve migration, branch pulmonary artery obstruction, pulmonary artery haemorrhage, and endocarditis.[49]

https://doi.org/10.1017/S1047951117002141 Published online by Cambridge University Press

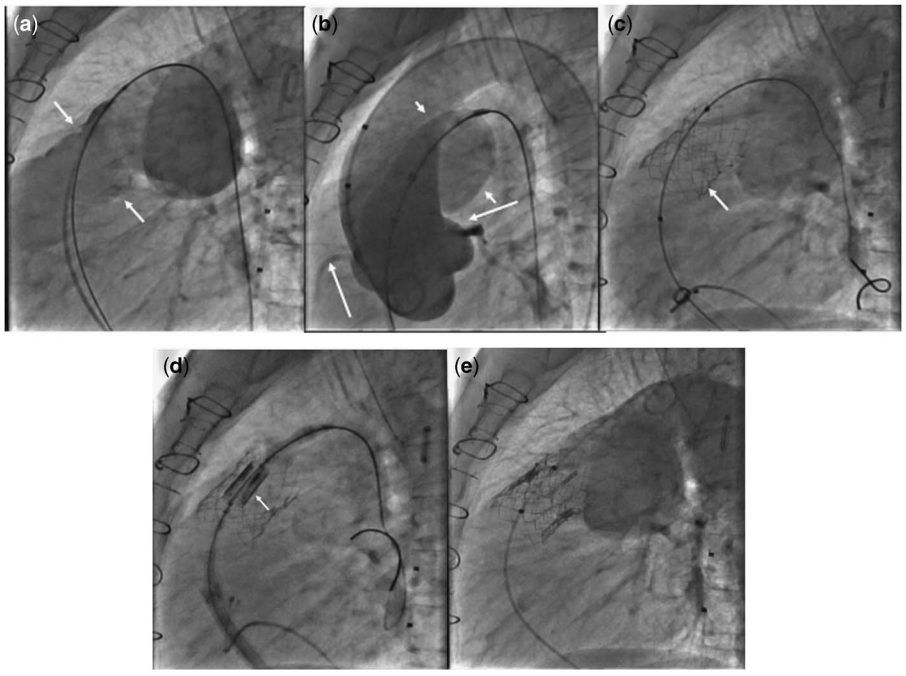

Figure 3.
*A 25-year-old female S/P tetralogy of Fallot with a conduit and severe pulmonary regurgitation underwent Edwards valve implantation. (**a**) Angiogram in the main pulmonary artery in the lateral projection demonstrating severe regurgitation. The two white arrows denote the boundary of the conduit. (**b**) Balloon sizing of the conduit during ascending aorta angiogram to assess proximity of coronary arteries from the right ventricle outflow tract. Two short white arrows denote the balloon in the right ventricle outflow tract/conduit and two long white arrows denote left and right coronary arteries with ample distance from balloon. (**c**) A P3110 Palmaz stent inflated to about 27 mm inside the conduit (arrow). (**d**) Cine fluoroscopy during positioning of a 26 mm Sapien valve (arrow) inside the landing zone (Palmaz stent). (**e**) Final angiogram in the main pulmonary artery after valve had been deployed showing no pulmonary regurgitation and good valve position.*

The future of cardiac catheterisation

Transcatheter pulmonary valve replacement for the native right ventricle outflow tract

The currently commercially approved valves can meet the needs of ~25% of patients with conduits and or bioprosthetic valves between the right ventricle and pulmonary artery. This leaves ~75% of patients who underwent transannular patch repair that are not suitable for these two valves. The Venus P valve (Venus Medtech) and the Medtronic Harmony valve (Medtronic Inc) have been developed for this specific patient population.

The Venus P valve. The Venus P valve (Venus Medtech, Shanghai, China) is a recently developed self-expanding transcatheter heart valve, designed to adapt to a dilated right ventricular outflow tract.[50,51] The valve consists of a stent of a Nitinol frame, and the valve leaflets are made of porcine pericardium preserved in low-concentration solutions of buffered gluteraldehyde and are hand-sewn to the multilevel self-expanding Nitinol frame. The frame has proximal and distal flares to anchor the valve in the right ventricular outflow tract and pulomonary artery bifurcation, respectively. The proximal flare is completely covered by pericardial tissue, whereas the distal flare is an open cell wire frame allowing access into the pulomonary artery branches (Fig 4). The middle part is tubular (straight) and is fully covered, housing the valve, and is intended to be expanded in the main pulmonary artery. For ease of identification, there are two radiopaque platinum markers at the proximal flare junction with the straight part and at the distal flare junction with the straight part. The valve is located about 5 mm distal to the proximal marker. The diameters and the lengths of the straight part range from 18 to 34 mm (in 2 mm increments) and from 20 to 35 mm (in 5 mm increment), respectively. After cardiac magnetic resonance and angiographic evaluation and in order to reduce the possibility of obstruction of the right ventricular body or pulomonary artery branches and to reduce paravalvar leak, the valve length can be selected to match the length of the main pulmonary artery. The proximal and distal flare diameters are 10 mm larger than the diameter of the straight segment. There are two small "ears" at the proximal part of the valve for attachment to the delivery system. The frame is made of a single Nitinol tube by lasers cutting. This design improves the frame integrity; however,

https://doi.org/10.1017/S1047951117002141 Published online by Cambridge University Press

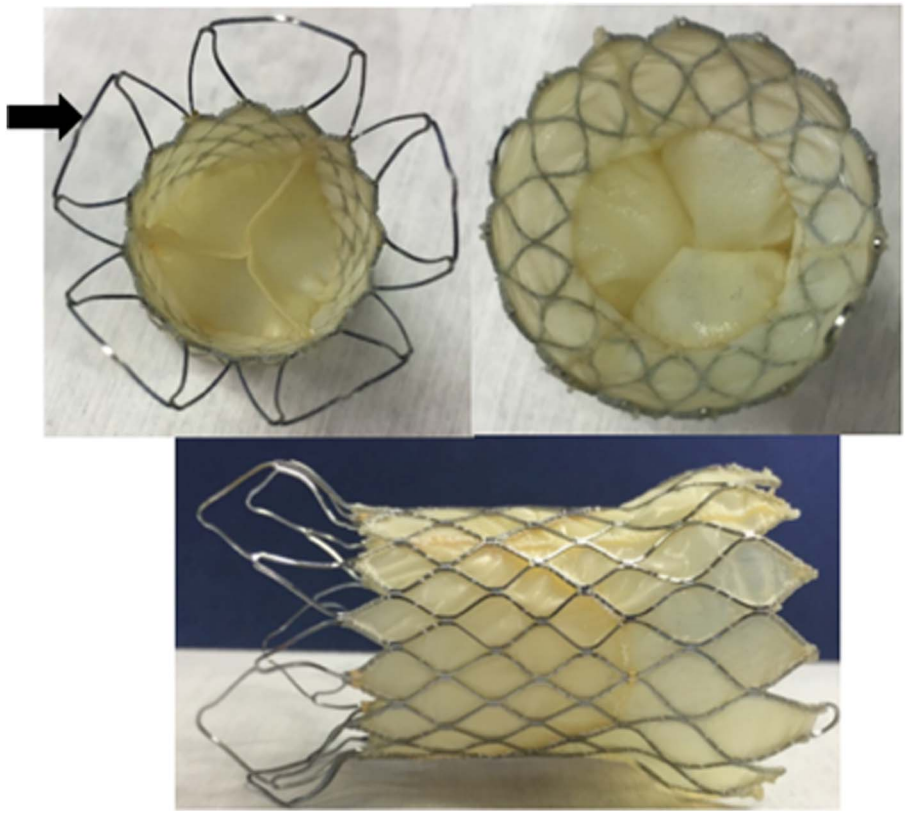

Figure 4.
Venus P valve. Note the open cells at the distal part of the stent (black arrow). These cells can be positioned at the bifurcation of the pulmonary arteries allowing access to these vessels.

manufacturing of the different sizes to fit the anatomy of all the patients is more time-consuming and costly. There are five – previously six – open cells in the distal flared part – to allow easy access to the branch pulomonary artery – with a wire across – to decrease the chance of fracture of the distal stent. The delivery system consists of a 20–22 Fr capsule and a 16 Fr, 100-cm-long shaft, with a rotating handle for deployment of the valve. The valve prosthesis is loaded into the capsule by submerging the Nitinol frame in sterilised cold saline solution and crimping the frame with a crimper provided by the manufacturer. Early experience with the valve has been very encouraging and the valve is undergoing a clinical trial for CE approval.

The Medtronic Harmony valve. It consists of a nitinol frame covered by a polyester cloth with valve leaflets made of porcine pericardium. The internal diameter is 22 mm. The valve requires a 25 Fr delivery sheath. The valve is undergoing an early feasibility clinical trial in North America with very encouraging early results.

Beijing Med-Zenith PT valve. Similar to the Venus P valve, it is a nitinol frame covered by porcine pericardial tissue with porcine pericardial leaflets. This valve is available in three sizes (20, 23, 26 mm) and requires 18 Fr delivery system. The valve will undergo a clinical trial soon.

Bioresorbable Stent Scaffold

Balloon-expandable stainless steel stents were developed by Palmaz in 1985[52,53] and since their first clinical use in CHD,[54] they have become the primary technical tool to treat vascular stenoses – peripheral pulmonary arterial stenosis and coarctation of the aorta and other systemic vessels. The major limiting factors using bare metallic stents include thrombosis and late in-stent stenosis, lack of growth as the child ages, and limited expansion potential. The development of drug-eluting stents, a significant breakthrough in stent technology for coronary applications – reducing restenosis rates,[55] has had limited application in CHD given the existing device diameters and systemic drug effects.

Thus, the concept of bioabsorbable stent has found traction as an alternative to bare metallic stents, both in adult and pediatric applications. Bioabsorbable stent has a number of advantages over existing bare metallic stents designs. Once bioabsorbed, they potentially leave behind a healed endothelialised natural vessel[55] with further growth potential. Stent

https://doi.org/10.1017/S1047951117002141 Published online by Cambridge University Press

thrombosis and in-stent luminal loss are not likely to occur as the stent is gone. Furthermore, bioabsorbable stent can act as a platform similar to their metal counterparts for either drugs or genes to promote vessel growth. Finally, bioabsorbable stent is readily compatible with MRI and CT imaging. Some animal trials have been conducted using stents with diameters of 6 mm in the aorta and branch pulmonary arteries. The initial data are encouraging. We are in discussion with the US Food and Drug Administration for an early feasibility study in pediatric patients. We are targeting the use of bioabsorbable stent in coarctation of the aorta and branch pulmonary arteries.

Should this technology prove useful in paediatric patients, the next steps will be designing a percutaneous valve made of large scaffold with leaflets made of stem cell technology. This will be the ultimate goal of treating paediatric patients with severe pulmonary regurgitation.

Percutaneous Fontan completion in the catheterisation laboratory

Among the many achievements of the late Gerd Hausdorf was the pre-conditioning of patients with single ventricle physiology who underwent the cavopulmonary shunt to prepare the scene for completion of their Fontan circuits in the catheterisation laboratory. As first step, a surgical hemi-Fontan procedure was carried out and a multifenestrated patch was inserted into the right atrium with subtotal banding of the junction between the atrium and superior caval vein. A few months later, Fontan completion was achieved by either stenting this junction – bare metallic stents – and closure of the fenestrations with the Rashkind umbrellas or by placing a covered stent from the inferior caval vein to the junction of the superior vena cava. The group successfully treated eight patients without any mortality.[56] A few other groups have achieved similar results since that report, including the group in Saudi Arabia, Toronto, and Columbus among others. The challenges of these procedures included the residual right to left shunt from the inferior limb of the covered stent graft – inferior caval vein-right atrium junction. This was due to stent-vessel separation during dilation of the stent. To treat this complication, another short-covered stent was implanted between the inferior vena cava and the inferior limb of the previously implanted stent covering the site of the leak. Further technical difficulties the authors faced were the inability to create vessel/chamber continuity – from the superior caval vein to the right atrium – due to force needed to perforate and dilate the area to an acceptable diameter. One potential solution that Evan M. Zahn, MD has been working on is the creation of a perforatable membrane (ePTFE) with a flat predetermined diameter stent. Another potential solution is to avoid intracardiac stent graft placement and work on an extracardiac tunnel concept. This work has been done by Zahid Amin, MD who proposed the idea that after completing the bidirectional Glenn anastomosis, place a tiny balloon expandable stent/wire mesh sewn at the inferior vena cava end of the Gore-Tex tube. The conduit is sewn in a trap-door fashion to the inferior vena cava and a flat stent placed on the pulmonary end of the tube. A few months later, a stent is placed from the inferior vena cava inside the Gore-Tex tube that connects the inferior vena cava with the pulmonary artery end.

Therefore, as we can see, Fontan completion in the catheterisation laboratory is feasible using different techniques. To achieve this successfully, a multicenter clinical trial should be initiated with a well-defined protocol. Availability of materials is essential for the success of this protocol.

Conclusion

Cardiac catheterisation for children/adults with CHD has progressed from the initial balloon atrial septostomy performed in the 1960s to balloon valvuloplasty and angioplasty in the 1980s and to septal defect closure in the 1990s and early 2000s to what we are currently doing of hybrid interventions and percutaneous pulmonary valve implantation. The future is very bright for this modality. The advent of bioresorbable stent technology is yet to be applied in CHD and, finally, Fontan completion in the catheterisation laboratory is the dream of every interventional cardiologist.

Acknowledgement

The authors wish to thank John P. Cheatham, MD, Evan Zahn, MD and Zahid Amin MD for providing information on their work.

Disclosure

Z.M.H. is a consultant for NuMED Inc; Occlutech, and Venus Medtech.

References

1. Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy: a palliative approach to complete transposition of the great arteries. JAMA. 1966; 196: 991–992.
2. Kan JS, White RI Jr, Mitchell SE, Gardner TJ. Percutaneous balloon valvuloplasty: a new method for treating congenital pulmonary-valve stenosis. N Engl J Med 1982; 307: 540–542.

3. Feltes TF, Bacha E, Beekman RH, et al. Indications for cardiac catheterization and intervention in pediatric cardiac disease: a scientific statement from the American Heart Association. Circulation 2011; 123: 2606–2652.
4. Hijazi ZM, Awad SM. Pediatric cardiac interventions. JACC Cardiovasc Interv 2008; 1: 603–611.
5. Lababidi Z. Aortic balloon valvuloplasty. Am Heart J 1983; 106: 751–752.
6. Qureshi SA, Redington AN, Wren C, et al. Recommendations of the British Pediatric Cardiac Association for therapeutic cardiac catheterization in congenital heart disease. Cardiol Young 2000; 10: 649–667.
7. Porstmann W, Wierny L, Warnke H. The closure of the patent arteriosus without thoracotomy. (preliminary report). Thoraxchir Vask Chir 1967; 15: 199–203.
8. Gatzoulis MA, Rodington AN, Somerville J, Shore DF. Should atrial septal defects in adults be closed? Ann Thorac Surg 1996; 61: 657–659.
9. Silvestry FE, Cohen MS, Armsby LB, et al. Guidelines for the echocardiographic assessment of atrial septal defect and patent foramen ovale: from the American Society of Echocardiography and Society for Cardiac Angiography and Interventions. J Am Soc Echocardiogr 2015; 28: 910–958.
10. Al-Hindi A, Cao QL, Hijazi ZM. Transcatheter closure of secundum atrial septal defect in the elderly. J Invasive Cardiol 2009; 21: 70–75.
11. Du ZD, Hijazi ZM, Kleinman CS, Silverman NH, Larntz K. Comparison between transcatheter and surgical closure of secundum atrial septal defect in children and adults: results of a multicenter non-randomized trial. J Amer Coll Cardiol 2002; 39: 1836–1844.
12. Holzer R, Balzer D, Cao QL, Lock K, Hijazi ZM. 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 Amer Coll Cardiol 2004; 43: 1257–1263.
13. Waight DJ, Bacha EA, Kahana M, Cao QL, Heitschmidt M, Hijazi ZM. Catheter therapy of Swiss Cheese ventricular septal defects using the Amplatzer MUSCULAR VSD occluder. Cath Cardiovasc Intervent 2002; 55: 355–361.
14. Fu YC, Bass J, Amin Z, et al. Transcatheter closure of perimembranous ventricular septal defects using the new Amplatzer membranous VSD occluder. J Amer Coll Cardiol 2006; 47: 319–325.
15. Holzer R, deGiovanni 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 Intervent 2006; 68: 620–628.
16. Bacha EA, Cao QL, Starr JP, Waight DJ, Ebeid MR, Hijazi ZM. Perventricular device closure of muscular ventricular septal defects on the beating heart: technique and results. J Thorac Cardiovasc Surg 2003; 126: 1718–1723.
17. Holzer R, Marshall A, Kreutzer J, et al. Hybrid procedures: adverse events and procedural characteristics-results of a multi-institutional registry. Congenit Heart Dis. 2010; 5: 233–242.
18. Mahle WT, Spray TL, Wernovsky G, et al. Survival after reconstructive surgery for hypoplastic left heart syndrome: a 15-year experience from a single institution. Circulation 2000; 10: III136–III141.
19. Stasik SN, Goldberg CS, Bove EL, et al. Current outcomes and risk factors for the Norwood procedure. J Thorac Cardiovasc Surg 2006; 131: 412–417.
20. Ohye RG, Sleeper LA, Mahony L, et al. Comparison of shunt types in the Norwood procedure for single-ventricle lesions. N Engl J Med 2010; 362: 1980–1992.
21. Sarajuuri A, Jokinen E, Puosi R, et al. Neurodevelopmental and neuroradiologic outcomes in patients with univentricular heart aged 5 to 7 years: related risk factor analysis. J Thorac Cardiovasc Surg 2007; 133: 1524–1532.
22. Gibbs JL, Rothman MT, Rees MR, et al. Stenting of the arterial duct: a new approach to palliation for pulmonary atresia. Br Heart J 1992; 67: 240–245.
23. Ruiz CE, Gamra H, Zhang HP, et al. Stenting of the ductus arteriosus as a bridge to cardiac transplantation in infants with the hypoplastic left-heart syndrome. N Engl J Med 1993; 328: 1605–1608.
24. Gibbs JL, Wren C, Watterson KG, et al. Stenting of the arterial duct combined with banding of the pulmonary arteries and atrial septectomy or septostomy: a new approach to palliation for the hypoplastic left heart syndrome. Br Heart J 1993; 69: 551–555.
25. Gibbs JL, Uzun O, Blackburn ME, et al. Fate of the stented arterial duct. Circulation 1999; 99: 2621–2625.
26. Akintuerk H, Michel-Behnke I, Valeske K, et al. Stenting of the arterial duct and banding of the pulmonary arteries: basis for combined Norwood stage I and II repair in hypoplastic left heart. Circulation 2002; 105: 1099–1103.
27. Akintuerk H, Michel-Behnke I, Valeske K, et al. Hybrid transcatheter-surgical palliation: basis for univentricular or biventricular repair: the Giessen experience. Pediatr Cardiol 2007; 28: 79–87.
28. Galantowicz M, Cheatham JP. Lessons learned from the development of a new hybrid strategy for the management of hypoplastic left heart syndrome. Pediatr Cardiol 2005; 26: 190–199.
29. Galantowicz M, Cheatham JP, Phillips A, et al. Hybrid approach for hypoplastic left heart syndrome: intermediate results after the learning curve. Ann Thorac Surg 2008; 85: 2063–2070.
30. Holzer RJ, Wood A, Chisolm JL, et al. Atrial septal interventions in patients with hypoplastic left heart syndrome. Cathet Cardiovasc Interv 2008; 72: 696–704.
31. Stoica SC, Philips AB, Egan M, et al. The retrograde aortic arch in the hybrid approach to hypoplastic left heart syndrome. Ann Thorac Surg 2009; 88: 1939–1946.
32. Naguib AN, Winch P, Schwartz L, et al. Anesthetic management of the hybrid stage 1 procedure for hypoplastic left heart syndrome (HLHS). Paediatr Anaesth 2010; 20: 38–46.
33. Luce WA, Schwartz RM, Beauseau W, et al. Necrotizing enterocolitis in neonates undergoing the hybrid approach to complex congenital heart disease. Pediatr Crit Care Med 2011; 12: 46–51.
34. Gafoor S, Zahn EM. Intraoperative stent implantation. In: Sievert H, Qureshi S, Wilson N, Hijazi ZM, (eds). Interventions in Structural, Valvular, and Congenital Heart Disease. Taylor and Francis Group, Boca Raton, FL, 2015: 777–791.
35. Bonhoeffer P, Boudjemline Y, Saliba Z, et al. Percutaneous replacement of pulmonary valve in a right-ventricle to pulmonary artery prosthetic conduit with valve dysfunction. Lancet 2000; 356: 1403–1405.
36. Malekzadeh-Milani S, Ladouceur M, Cohen S, Iserin L, Boudjemline Y. Results of transcatheter pulmonary valvulation in native or patched right ventricular outflow tracts. Arch Cardiovasc Dis 2014; 107: 592–598.
37. O'Byrne ML, Glatz AC, Mercer-Rosa L, et al. Trends in pulmonary valve replacement in children and adults with tetralogy of fallot. Am J Cardiol 2015; 115: 118–124.
38. Quail MA, Frigiola A, Giardini A, et al. Impact of pulmonary valve replacement in tetralogy of Fallot with pulmonary regurgitation: a comparison of intervention and nonintervention. Ann Thorac Surg 2012; 94: 1619–1626.
39. Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010; 31: 2915–2957.
40. Silversides CK, Kiess M, Beauchesne L, et al. Canadian Cardiovascular Society 2009 Consensus Conference on the management of adults with congenital heart disease: outflow tract obstruction,

coarctation of the aorta, tetralogy of Fallot, Ebstein anomaly and Marfan's syndrome. Can J Cardiol 2010; 26: e80–e97.
41. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to develop guidelines on the management of adults with congenital heart disease). Circulation. 2008; 118: e714–e833.
42. Lee C, Kim YM, Lee CH, et al. Outcomes of pulmonary valve replacement in 170 patients with chronic pulmonary regurgitation after relief of right ventricular outflow tract obstruction: implications for optimal timing of pulmonary valve replacement. J Am Coll Cardiol 2012; 60: 1005–1014.
43. Brown DW, McElhinney DB, Araoz PA, et al. Reliability and accuracy of echocardiographic right heart evaluation in the U.S. Melody Valve Investigational Trial. J Am Soc Echocardiogr 2012; 25: 383–392.
44. Cheatham SL, Holzer RJ, Chisolm JL, Cheatham JP. The Medtronic Melody(R) transcatheter pulmonary valve implanted at 24-mm diameter – it works. Cathet Cardiovasc Interv 2013; 82: 816–823.
45. 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: 535–542.
46. Kenny D, Hijazi ZM, Kar S, et al. Percutaneous implantation of the Edwards SAPIEN transcatheter heart valve for conduit failure in the pulmonary position: early phase 1 results from an international multicenter clinical trial. J Am Coll Cardiol 2011; 58: 2248–2256.
47. Butera G, Milanesi O, Spadoni I, et al. Melody transcatheter pulmonary valve implantation. Results from the registry of the Italian Society of pediatric cardiology. Catheter Cardiovasc Interv 2013; 81: 310–316.
48. Zahn EM, Hellenbrand WE, Lock JE, McElhinney DB. Implantation of the melody transcatheter pulmonary valve in patients with a dysfunctional right ventricular outflow tract conduit early results from the U.S. Clinical trial. J Am Coll Cardiol 2009; 54: 1722–1729.
49. Sharma A, Cote AT, Hosking MCK, Harris KC. A systematic review of infective endocarditis in patients with bovine jugular vein valves compared with other valve types. JACC Cardiovasc Interv 2017; 10: 1449–1458.
50. Cao QL, Kenny D, Zhou D, et al. Early clinical experience with a novel self-expanding percutaneous stent-valve in the native right ventricular outflow tract. Catheter Cardiovasc Interv 2014; 84: 1131–1137.
51. Promphan W, Prachasilchai P, Siripornpitak S, Qureshi SA, Layangool T. Percutaneous pulmonary valve implantation with the Venus P-valve: clinical experience and early results. Cardiol Young 2016; 26: 698–710.
52. Palmaz JC, Sibbitt RR, Reuter SR, Garcia F, Tio FO. Expandable intrahepatic portocaval shunt stents: early experience in the dog. Am J Roentgenol 1985; 145: 821–825.
53. Palmaz JC, Sibbitt RR, Tio FO, Reuter SR, Peters JE, Garcia F. Expandable intraluminal vascular graft: a feasibility study. Surgery 1986; 99: 199–205.
54. O'Laughlin MP, Perry SB, Lock JE, Mullins CE. Use of endovascular stents in congenital heart disease. Circulation 1991; 83: 1923–1939.
55. Serruys PW, Ormiston JA, Onuma Y, et al. A bioabsorbable everolimus-eluting coronary stent system (ABSORB): 2-year outcomes and results from multiple imaging methods. Lancet. 2009; 373: 897–910.
56. Hausdorf G, Schneider M, Konertz W. Surgical preconditioning and completion of total cavopulmonary connection by interventional cardiac catheterization: a new concept. Heart 1996; 75: 403–409.