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Review

Percutaneous Valve Interventions in the Adult Congenital Heart Disease Population: Emerging Technologies and Indications

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ABSTRACT

Adult survivors with congenital heart disease are not cured and residual cardiac valve lesions are common and contribute substantially to long-term morbidity. Given the increased risk of reoperations in patients with previous cardiac surgery, percutaneous treatment options have been developed. Initially percutaneous therapies focused on right ventricular outflow tract lesions, but they have now expanded to include mitral and aortic valve interventions. Although some of these procedures, such as balloon valvuloplasty of pulmonary valve stenosis and percutaneous pulmonary valve replacement, have become standard of care, there are many new and evolving technologies that will likely become important treatment strategies over the coming decade. The key for success of these transcatheter valve procedures is the careful evaluation of the patient's individual anatomy

RÉSUMÉ

Les patients atteints d'une cardiopathie congénitale qui survivent jusqu'à l'âge adulte ne sont pas guéris; ils présentent souvent des lésions valvulaires cardiaques résiduelles qui augmentent considérablement la morbidité à long terme. En raison du risque accru de réopération chez les patients ayant déjà subi une chirurgie cardiaque, des techniques de traitement percutané ont été mises au point. Initialement, les traitements percutanés visaient surtout les lésions des voies d'éjection du ventricule droit, mais leur portée s'est maintenant élargie et englobe des interventions touchant la valve mitrale et la valve aortique. Bien que certaines de ces interventions, comme la valvuloplastie par ballonnet en cas de sténose de la valve pulmonaire ou le remplacement valvulaire pulmonaire par voie percutanée, fassent maintenant partie des soins couramment utilisés, beaucoup de

Given the success of modern heart surgery and improved cardiology care, congenital heart disease has been transformed from a once deadly condition with high rates of childhood mortality into a treatable condition, with survival to adulthood having become the rule for the vast majority of affected patients, even those with complex disease.[1] This has led to rapidly expanding cohorts of adult survivors with congenital heart disease.[2] These patients are not cured.[3] Numerous studies have demonstrated a shift of morbidity and mortality from childhood to adulthood, often affecting these adults at young and middle ages.[1,4-7] In these populations, residual valve lesions are common and contribute substantially to long-term morbidity. Valve lesions in particular increase the risk of heart failure and arrhythmias. Given the increased risk of reoperations in patients with previous cardiac surgery, innovative interventionalists and engineers have created percutaneous treatment options for residual valve lesions over the past few decades.[8]

The aim of the present article is to provide a general overview of common valve lesions in the adult congenital heart disease population, indications for intervention, and the role of percutaneous treatment options as part of the long-term management strategy. We also provide an outlook on evolving and emerging technologies that may considerably broaden our therapeutic armamentarium in the future.

General Considerations

Adults with congenital heart disease—repaired, unrepaired or palliated—represent a unique cohort of complex patients in adult cardiology. To improve outcomes, these patients require multidisciplinary specialist care during their entire adult lives.[9] This requires a thorough understanding of the individual

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and physiology and a multidisciplinary assessment involving cardiologists specialized in adult congenital heart disease, specialized imagers, cardiac surgeons, and interventionalists. Because many of these percutaneous interventions are relatively new, long-term outcomes are not yet well defined, dictating the need for careful and structured long-term observational studies on outcomes of these novel procedures, which will allow refining the indications of a specific intervention and to improve its technical aspects. The aim of this article is to provide an overview of common valve lesions in the adult congenital heart disease population and to discuss treatment options and strategies with a specific focus on percutaneous options.

techniques nouvelles et en évolution occuperont vraisemblablement une place importante dans l'arsenal thérapeutique au cours de la prochaine décennie. La réussite de telles interventions valvulaires par cathétérisme interventionnel passe par l'évaluation minutieuse des caractéristiques anatomiques et physiologiques de chaque patient et par une examenétude de cas par une équipe multidisciplinaire formée de cardiologues spécialisés en cardiopathie congénitale chez l'adulte, de spécialistes en imagerie, de chirurgiens cardiaques et de cardiologues interventionnels. Comme beaucoup de ces interventions percutanées sont relativement nouvelles, on n'en connaît pas encore bien les résultats à long terme, d'où la nécessité de réaliser des études observationnelles rigoureuses et structurées de longue durée qui permettront de mieux cibler les indications d'une intervention particulière et d'en améliorer les aspects techniques. Le présent article a pour but de donner un aperçu des lésions valvulaires fréquemment observées dans la population d'adultes atteints de cardiopathie congénitale et de présenter les options et stratégies thérapeutiques, en mettant l'accent sur les interventions percutanées.

patient's anatomy and (patho)physiology (Fig. 1) to tailor decision making, optimize treatment options and planning of interventions, and anticipate the subsequent changes in the patient's hemodynamic profile.

Whereas decision making in the symptomatic patient is often relatively straight forward, decision making in the asymptomatic patient is more challenging, given the poor evidence base for "prophylactic" valve interventions and the absence of randomized prospective studies in the field of adult congenital heart disease. Because many of these young adults are expected to survive for many decades, competing risks, such as subsequent reinterventions, infective endocarditis, the need for anticoagulation, and the risk of pregnancy, should be considered in the indication for, and timing of, a valve intervention.[10]

Interventions on the Pulmonary Valve

Although pulmonary valve dysfunction is rare in patients with acquired heart disease, the management of right ventricular outflow tract lesions remains center-stage in congenital heart disease.

Isolated severe pulmonary valve stenosis secondary to congenitally malformed pulmonary valves (ie, bicuspid valves) is usually treated in childhood, and therefore significant native pulmonary valve stenosis is less common in the adult congenital cardiac population. However, adults may present with unrepaired pulmonary stenosis or residual pulmonary valve stenosis after a childhood intervention. Valve interventions are recommended in symptomatic patients with severe pulmonary valve stenosis and may be considered in asymptomatic patients with severe stenosis. Many decades ago, balloon valvuloplasty became accepted as first-line treatment in children and adults with isolated pulmonary valve stenosis, with excellent long-term results.[11,12] Surgical valve replacement with a bioprosthetic valve is usually reserved for treatment of infundibular obstruction or pulmonary annular hypoplasia.

Pulmonary regurgitation is a common valve lesion in the adult congenital cardiac population. It is often found as a residual lesion in adults who had undergone surgical pulmonary valvotomy in childhood, and residual pulmonary regurgitation is the most common hemodynamic residual lesion in adults with repaired tetralogy of Fallot. Tetralogy of Fallot is the most common cyanotic heart defect, affecting about 1 in 2500 live births.[13] Surgical repair in tetralogy of Fallot typically includes patch closure of the ventricular septal defect and relief of right ventricular outflow tract obstruction. Based on observations of long-term outcomes in adulthood, there has been a remarkable evolution in the surgical repair strategies of tetralogy of Fallot over the past few decades. Initial intracardiac repair often involved a transventricular approach and included a generous transannular right ventricular outflow tract patch to completely alleviate any outflow tract obstruction. However, this approach was eventually modified because transannular patch reconstruction disrupts the integrity of the pulmonary valve and results in severe pulmonary regurgitation with right ventricular dilation or systolic dysfunction. In contemporary cohorts, repair with the use of a transatrial approach, avoiding ventriculotomy scars, and right ventricular outflow tract reconstruction avoiding a transannular patch is preferred if the anatomy of an individual patient allows. Preserving the pulmonary valve, also known as the valve-sparing tetralogy of Fallot repair, prevents free pulmonary valve regurgitation, although some patients may be left with some degree of residual right ventricular outflow tract obstruction.

Symptomatic patients with moderate or severe pulmonary regurgitation with associated right ventricular dilation or dysfunction require pulmonary valve replacement.

The management of the asymptomatic patient with severe pulmonary regurgitation is more controversial, particularly the timing of pulmonary valve replacement in asymptomatic patients with repaired tetralogy of Fallot who had residual pulmonary regurgitation. Although there have been a number of studies showing reverse remodelling of the right ventricle and improvements in some cardiac outcomes after pulmonary valve replacement, the precise timing of pulmonary valve intervention is debated among experts. Indeed, a recent multicentre outcome study contradicted previous studies and

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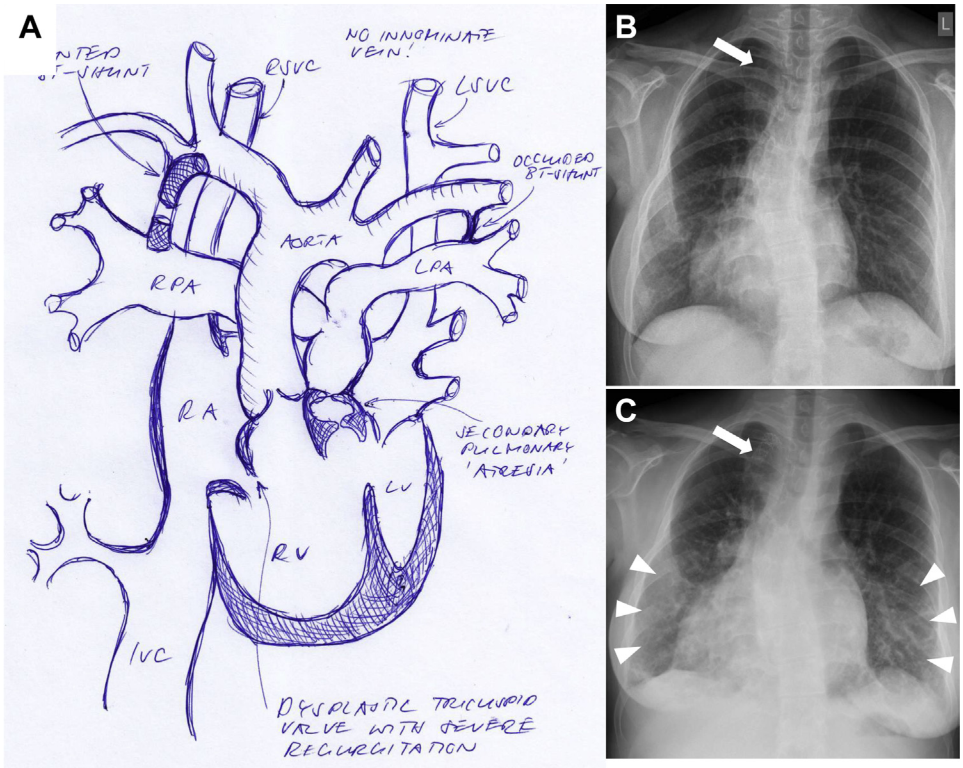

Figure 1. A 32-year old patient with complex cyanotic congenital heart disease. (**A**) Schematic diagram of cardiac and extracardiac anatomy depicting the right side Blalock-Taussig shunt as the sole source of pulmonary blood flow. Chest c-ray (**B**) before and (**C**) after dilatation and stenting of the obstructed right-side Blalock-Taussig shunt (**arrow**) for progressive cyanosis. The increased volume load of the right single ventricle after stenting of the Blalock-Taussig shunt led to worsening systemic tricuspid regurgitation with subsequent decompensated heart failure, pleural effusions, and pulmonary congestion (**arrowheads**). This case highlights the importance of anticipating hemodynamic changes before any percutaneous interventions in patients with complex congenital heart disease.

raised doubts on the notion that early and liberal prosthetic pulmonary valve replacement improved long-term outcomes in this patient population.[10,14-16] The liberal indication for pulmonary valve replacement for patients with repaired tetralogy of Fallot and residual severe pulmonary regurgitation has, over the past 2 decades, rapidly expanded the cohort of patients with pulmonary valve prosthesis and pulmonary valve conduits.[17] The experience with mechanical prosthesis in pulmonary position is very limited, because these implants are hampered by a relatively high risk of thromboembolic complications.[18] Therefore, at almost all centres the use of bioprosthetic valves (xenografts or homografts) has become the standard operation for right ventricular outflow tract lesions. All bioprosthetic valves eventually fail and require redo intervention for restoration of right ventricular outflow tract function.

Right ventricle—to—pulmonary artery conduits are required for primary intracardiac repair in patients with tetralogy of Fallot and significant pulmonary hypoplasia/atresia, complete transposition of the great arteries, ventricular septal defects requiring the Rastelli operation, and truncus arteriosus communis. Severe symptomatic conduit stenosis is an indication for repair. Although balloon valvuloplasty of stenotic bioprosthetic conduits may offer short-term amelioration of valve function, it was the seminal work of Philipp Bonhoeffer and his teams that paved the way to a revolutionary concept in interventional cardiology: the percutaneous replacement of heart valves.[19] After refinement of the technique with the use of pre-stenting of the right ventricular outflow tract and paying particular attention to potential compression of coronary arteries, percutaneous replacement of failing conduits and later bioprosthetic valves in pulmonary position with the Melody valve system (Medtronic Inc, Minneapolis, MN) has emerged as the standard treatment option for patients with suitable anatomy (Fig. 2).[20,21] Given the limited upper diameter of the Melody system, other systems, such as the Sapien valve (Edwards Lifesciences, Inc, Irvine, CA), have subsequently been successfully used for pulmonary valve replacement in patients with larger conduits exceeding the maximal diameter of the Melody system.[22]

A challenge remains in the interventional implantation of prosthetic valves in the native or patched right ventricular outflow tract that have dilated or distorted outflow tract anatomy.[23] The main challenge with currently available percutaneous systems remains the establishment of a stable “landing zone” for subsequent implantation of the stent-based percutaneous valve. In patients with suitable anatomy and diameter of the native or patched right ventricular outflow tract, direct implantation with or without pre-stenting has proved feasible (Fig. 3).[24,25] However, more than two-thirds of patients after a transannular patch repair have a right ventricular outflow tract that is too large for presently available

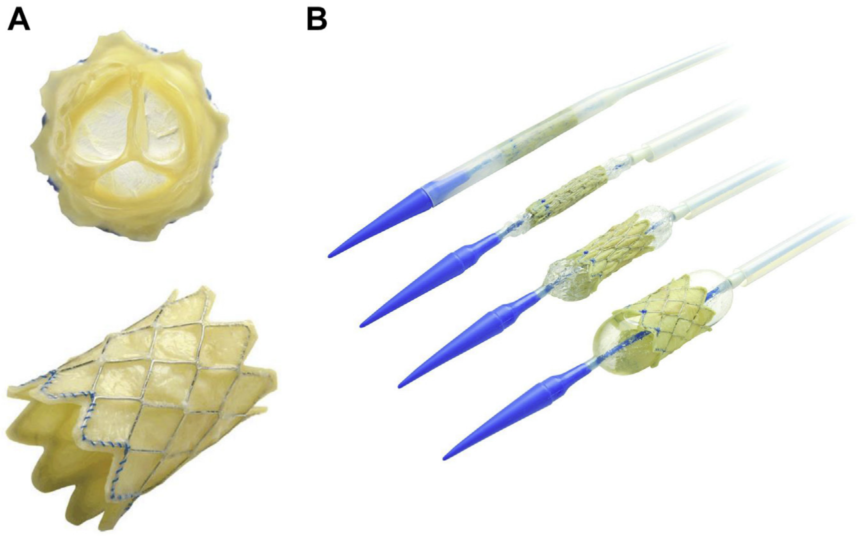

Figure 2. Transcatheter Melody valve (Medtronic Inc, Minneapolis, MN) for percutaneous replacement of pulmonary valves. (**A**) Stented percutaneous valve prosthesis. (**B**) Delivery system and assembly of the percutaneous valve on the delivery system. Reproduced with permission from Medtronic.

percutaneous valves. To address that deficiency a number of device companies have completed early feasibility and pivotal clinical studies investigating valves designed specifically for these large patulous outflow tracts.[26-28]

Although percutaneous replacement of the pulmonary valve has been a milestone in the management of right ventricular outflow tract lesions in patients with congenital heart disease, some caveats remain. These include a careful assessment of the procedural risk, which includes, as mentioned above, compression of coronary arteries and conduit rupture (Fig. 4). In addition, there has been the observation of a high rate of infective endocarditis after percutaneous pulmonary valve replacement.[29,30] In this regard, there are substantial differences in the rates of infective endocarditis between centers and potentially between different types of prosthetic valves.[31] It will be crucially important to delineate—and eliminate—risk factors for infective endocarditis in the future to pursue the success story of percutaneous pulmonary valve replacement.[32] The implantation of percutaneous pulmonary valves in patients with previous infective endocarditis remains contentious and should be evaluated on an individual basis after careful consideration of potentials risks and benefits.

Transcatheter aortic valve procedures

Bicuspid and unicuspid aortic valve disease accounts for most cases of significant aortic stenosis and regurgitation in the adult congenital cardiac clinic. Congenital aortic valve disease can occur in conjunction with other cardiac lesions. For example, patients with bicuspid aortic valve disease may also have coarctation of the aorta, highlighting the need for careful and complete cardiac evaluation in patients with congenital aortic stenosis. Bicuspid valve disease is also associated with aortopathy and, if the aorta is significantly dilated, may require surgical replacement at the time of valve surgery.

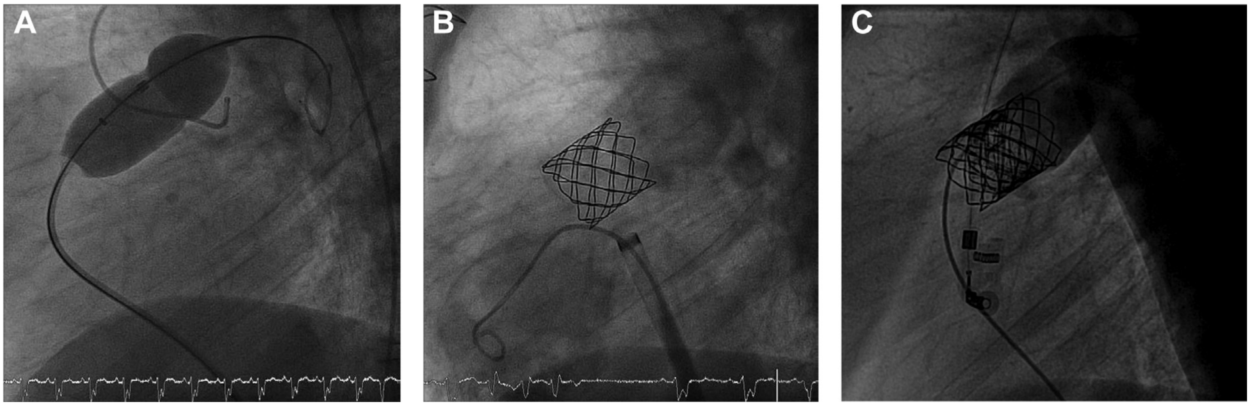

Figure 3. Percutaneous pulmonary valve replacement in a patient with patched right ventricular outflow tract. (**A**) Balloon sizing of the right ventricular outflow tract. (**B**) Pre-stenting of the right ventricular outflow tract to achieve a stable landing zone for the valved stent. (**C**) Implantation of a percutaneous pulmonary valve, 3 months after pre-stenting, with a 26 mm Sapien valve (Edwards Lifesciences, Inc, Irvine, CA).

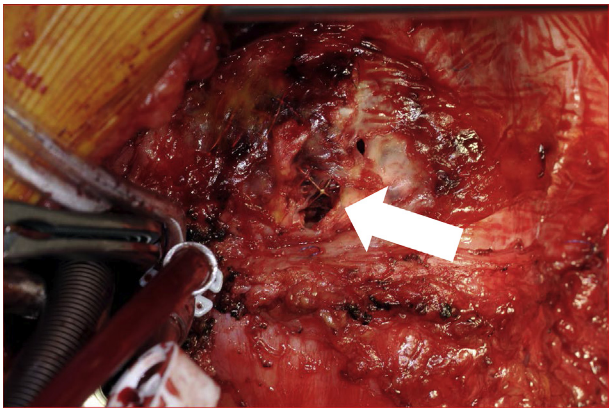

Figure 4. Ruptured homograft conduit after pre-stenting in preparation of percutaneous pulmonary valve replacement. The **arrow** points to the site of rupture of the pulmonary homograft at the time of surgical replacement.

Indications for aortic valve surgery for congenital aortic stenosis or regurgitation are similar to those for tricuspid aortic valve stenosis and regurgitation. Patients with severe congenital aortic stenosis who are symptomatic (class I recommendation), have a left ventricular ejection fraction < 50% (class I recommendation), or have abnormal exercise testing (class IIa recommendation) require valve intervention. Similarly, patients with severe aortic regurgitation who are symptomatic (class I recommendation), have a left ventricular ejection fraction < 50% (class I recommendation), or have significant left ventricular dilation (left ventricular end systolic dimension > 50 mm; class IIa recommendation) require valve replacement.

Surgical valve replacement is the standard approach to patients with congenital aortic stenosis or regurgitation. Young adults with congenital aortic stenosis or regurgitation who require surgery should be well informed about the various surgical options. Mechanical valves have longevity, but some young adults may prefer a bioprosthetic valve if anticoagulation is not desired or if they are women of childbearing age and wish to minimize pregnancy risks. The Ross procedure, at least in the hands of experienced surgeons, may offer longer freedom from reoperation than simple bioprosthetic aortic valve replacement. Valve repair for severe aortic regurgitation, often in conjunction with an aortic root replacement (David operation), may be a feasible option in selected cases.

Balloon dilation of the aortic valve is an accepted palliation for infants, children and adolescents with aortic valve stenosis, typically in the setting of a bicuspid or, less commonly, a unicuspid aortic valve. Although there is often some degree of postprocedural aortic regurgitation, balloon dilation allows avoidance of surgery until regurgitation becomes the predominant clinical concern and surgical approaches can be used. In adults with calcified aortic valve stenosis, the results of balloon valvuloplasty are much less favourable and this approach is rarely used. Even after successful reduction of systolic gradients, the effect is typically of very limited duration.[33]

As with the percutaneous options for pulmonary valve replacement, transcatheter aortic valve replacement has revolutionized the treatment of aortic valve stenosis in adults. Although surgical aortic valve replacement remains the standard treatment option in most young adults, percutaneous options may be a suitable alternative in selected young adult patients with congenital aortic stenosis and high perioperative risks (see case vignette 1, below). Careful multidisciplinary assessment of such patients and individualized decision making within the heart team is crucial for optimal patient selection and treatment success.

Of great interest within the field of congenital heart disease was the question whether transcatheter aortic valve replacement may be an option for patients with aortic valve stenosis caused by degeneration of bicuspid aortic valves. Although the early experience suggested higher procedural failure rates in patients with bicuspid aortic valves, these problems seem to have been overcome by new-generation devices and more accurate preprocedural identification of unfavourable anatomic valve characteristics.[34-36]

Case vignette 1: Percutaneous aortic valve replacement in a young woman

A 32-year-old female patient, born with Shone complex presented with heart failure due to severe prosthetic aortic valve regurgitation (21 mm Medtronic Freestyle prosthesis; Medtronic Inc), less than 2 years after the operation: She had a long surgical history, with aortic coarctation repair via thoracotomy and subsequently a total of 4 operations and reoperations for fibromuscular subaortic stenosis and aortic valve repair, all via sternotomy. At presentation, she had severe pulmonary hypertension and was in New York Heart Association (NYHA) functional class III-IV. After careful consideration of all therapeutic options, percutaneous aortic valve replacement was performed with placement of the aortic valve within the left ventricular outflow tract (Fig. 5) avoiding obstruction of coronary arteries. The patient had an uneventful periinterventional clinical course and was discharged home the second day after the intervention. She made a rapid and complete recovery with normalization of pulmonary pressures, normalization of biventricular function, and regression of secondary mitral valve regurgitation. More than 7 years after the intervention, prosthetic aortic valve function remained favourable with mild prosthetic valve stenosis, and the patient remained in NYHA functional class I.

Transcatheter mitral valve procedures

Isolated congenital mitral valve stenosis is rare in the adult population. Congenital mitral stenosis can occur as part of Shone syndrome, a congenital cardiac complex consisting of multiple left-side outflow tract obstructive lesions. Left atrioventricular valve stenosis can also occur after intracardiac repairs, such as a cleft mitral valve repair in patients with atrioventricular septal defects. Valve interventions are recommended in symptomatic patients with severe mitral stenosis. Compared with interventions in patients with rheumatic mitral valve disease, balloon dilation in patients with congenital dysplastic mitral valves are typically less effective, largely depending on anatomic characteristics of the failing mitral valves. Mitral stenosis physiology can be seen with cor

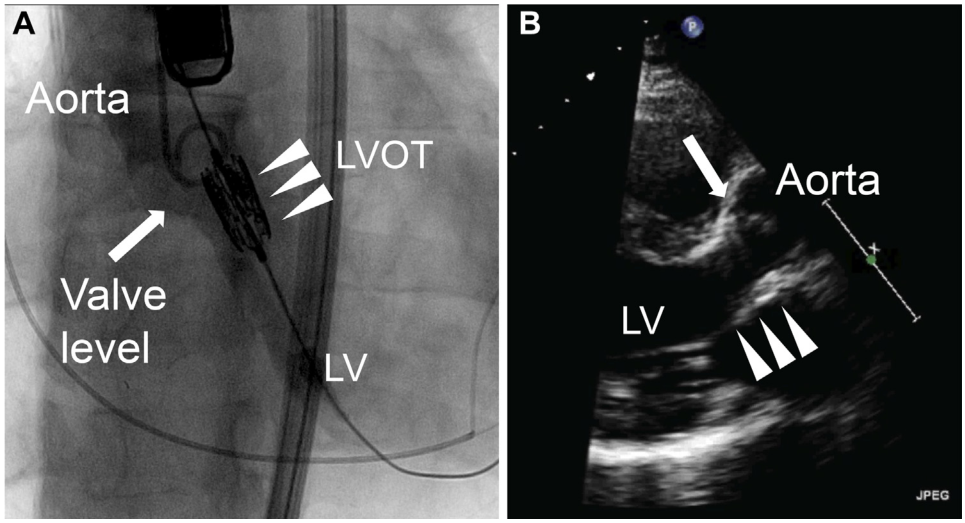

Figure 5. Implantation of a percutaneous Sapien valve (Edwards Lifesciences, Inc, Irvine, CA) within the left ventricular (LV) outflow-tract. (**A**) Fluoroscopy at the time of deployment of the Sapien valve. The **arrow** points to the level of the annulus of previously implanted Freestyle bioprosthesis; **arrowheads** point to the left ventricular outflow tract (LVOT). (**B**) Parasternal long-axis view after implantation of the Sapien valve within the LVOT (**arrowheads**) below the level of the Freestyle bioprosthesis (Medtronic Inc, Minneapolis, MN) (**arrow**).

triatriatum or supramitral rings, and these lesions require more complex surgical repairs.

Congenital mitral regurgitation most commonly occurs in the setting of myxomatous mitral valve disease with mitral valve prolapse or after left atrioventricular valve repairs in patients with atrioventricular septal defects. Patients with myxomatous mitral valve disease often have mitral anatomy that is suitable for repair. Repair of a cleft mitral valve is much more complex: The valve anatomy is not usually suitable for repair and valve replacement is more commonly required.

Percutaneous techniques for reduction of mitral regurgitation in patients with functional or degenerative mitral valve disease (eg, flail leaflets) are at present rarely used in patients with congenital heart disease (such as left atrioventricular valve regurgitation after repair of atrioventricular septal defects) owing to unsuitable valve leaflet anatomy.

Transcatheter tricuspid valve procedures

The tricuspid valve is anatomically the most complex and variable of all 4 heart valves. In congenital heart disease, the tricuspid valve can be the atrioventricular valve of the subpulmonary ventricle, the subaortic ventricle or guarding the atrioventricular junction of a single ventricle. Furthermore, in our experience, tricuspid valve function is particularly dependent on changes in hemodynamics and changes in (right ventricular) pre- and afterload. In the setting of significant tricuspid regurgitation, true systolic function of the right ventricular myocardium is difficult to assess, and simple measures such as right ventricular ejection fraction may overestimate the true contractility and contractile reserve of the right ventricular myocardium. The latter aspect may explain the relatively high risk of perioperative mortality in such patients, because the "true" right ventricular dysfunction may become apparent only after restoration of tricuspid valve competence.[37]

Ebstein anomaly is the most common congenital tricuspid valve lesion. It is characterized by failure of appropriate delamination of the tricuspid valve tissue from right ventricular myocardium during the embryologic development of the heart, which leads to the typical apical displacement of the tricuspid valve, hypoplasia of the septal and/or posterior leaflets, elongation of the anterior leaflet, and varying degrees of tricuspid regurgitation. Less commonly, there is tricuspid valve stenosis. Surgical repair of the valve is complex. In the current era, the cone reconstruction is the surgical repair of choice. When not feasible, valve replacement is an alternative. Percutaneous approaches are not used for this complex valve lesion.

In patients with tricuspid valve regurgitation in the setting of congenital heart disease, careful assessment of valve anatomy and the exact mechanism(s) of valve regurgitation is key to optimal planning of the therapeutic strategies. Comprehensive evaluation requires the application of multiple modalities, including echocardiography, magnetic resonance imaging, and cardiac catheterization. Before a surgical or interventional procedure, every effort should be made for optimization of hemodynamics, which may have a substantial impact in the case of functional tricuspid regurgitation (Fig. 6).

Functional tricuspid regurgitation, secondary to right ventricular dilation, is much more common in the adult congenital cardiac population. For example, patients with tetralogy of Fallot and severe right ventricular dilation often have functional tricuspid regurgitation. For patients with other indications for cardiac surgery, functional tricuspid regurgitation is often addressed at the time of surgery. However, given the risks of isolated surgery on the tricuspid valve, efforts are currently being made to develop interventional treatment options for treatment of functional tricuspid valve regurgitation. Figure 7 summarizes some of the currently available investigational devices and their relative frequency of use among such procedures as collected

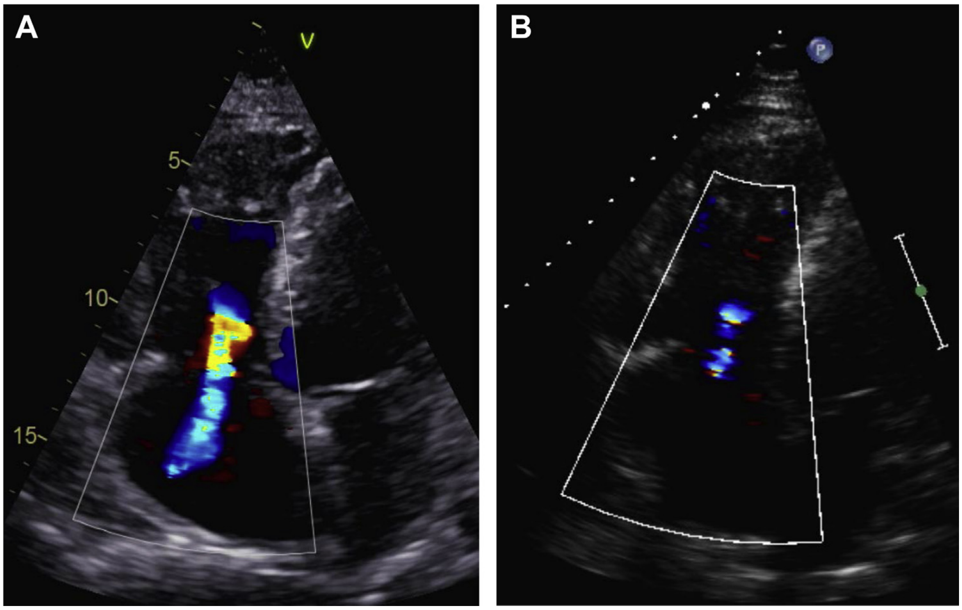

Figure 6. Regression of tricuspid valve regurgitation after implantation of a percutaneous pulmonary valve in a patient with progressive prosthetic pulmonary valve stenosis and secondary progressive tricuspid valve regurgitation. Apical view with color Doppler on the tricuspid valve depicting (**A**) moderate functional tricuspid regurgitation before and (**B**) mild tricuspid regurgitation after relief of right ventricular outflow tract obstruction.

in an international registry for interventional therapy of tricuspid regurgitation.[38] Currently, there are no data that allow formulating recommendations on when to consider transcatheter tricuspid valve repair in patients with congenital heart disease and functional tricuspid regurgitation. Decision making for use of such devices has to be individualized on a case-by-case basis with careful discussions within a dedicated heart team. Given the importance of tricuspid valve function on the long-term outcomes in adults with congenital heart disease, careful evaluation of the results of these ongoing studies and registries is important, and devices which may be of interest should be further investigated in patients with congenital heart disease.

Valve-in-valve procedures

The rapid evolution of techniques for percutaneous valve replacement in patients with degenerated bioprosthetic conduits or valves will likely lead to an expansion of the use of bioprosthetic valves and conduits at the time of initial valve surgery in patients who would traditionally meet the indication for mechanical prosthetic valve replacement. Although valve-in-valve percutaneous options for the failing pulmonary valve prosthesis have become standard of care in many instances, with relatively predictable outcomes, the performance and risk of valve-in-valve options for a bioprosthesis in aortic position depends on the type of bioprosthesis and requires careful assessment to avoid patient-prosthesis mismatch.[39] Long-term outcome data of such procedures are lacking. This has to be carefully discussed with patients when a decision for the choice of aortic valve prosthesis is made.

Valve-in-valve or valve-in-ring procedures may be an option for failing bioprostheses or ring annuloplasties in the mitral or tricuspid position, particularly for patients at high risk of reoperation.[40,41] In patients with congenital heart disease, the option of increasing the durability of a surgically implanted tricuspid valve bioprosthesis is particularly encouraging in patients with Ebstein anomaly, in whom surgical valve repair was not possible. First data from an international registry show promising results of using a valve-in-valve strategy in such patients.[42]

Paravalvular leaks and pseudoaneurysms

Paravalvular leaks present a unique challenge after prosthetic heart valve implantations. Such leaks in the setting of infective endocarditis usually require urgent surgical repair or replacement of the affected valve, which carries a high perioperative risk. In the absence of an infectious process, paravalvular leaks can lead to heart failure or hemolysis, depending on the size and localization of the defect(s). Given the typically high risk of reoperations in this setting, an interdisciplinary evaluation of percutaneous treatment options should always be considered, particularly in patients with multiple previous open heart surgeries, as is often the case in adults with congenital heart disease (see case vignette 2).

Case vignette 2: Percutaneous closure of a large pseudoaneurysm

A patient with bicuspid aortic valve and severe aortic stenosis underwent replacement of the aortic valve with a bioprosthesis at the age of 32 years. At the age 41 years he was successfully treated for infective endocarditis but had a second episode 1 year later, at the age of 42 years. He had a large vegetation and partial dehiscence of the aortic valve bioprosthesis, requiring urgent surgical valve replacement. The surgical procedure was technically challenging, requiring replacement of the entire aortic root with a biocomposite graft and reimplantation of the coronary arteries. Early after the operation a small dehiscence of the proximal graft anastomosis

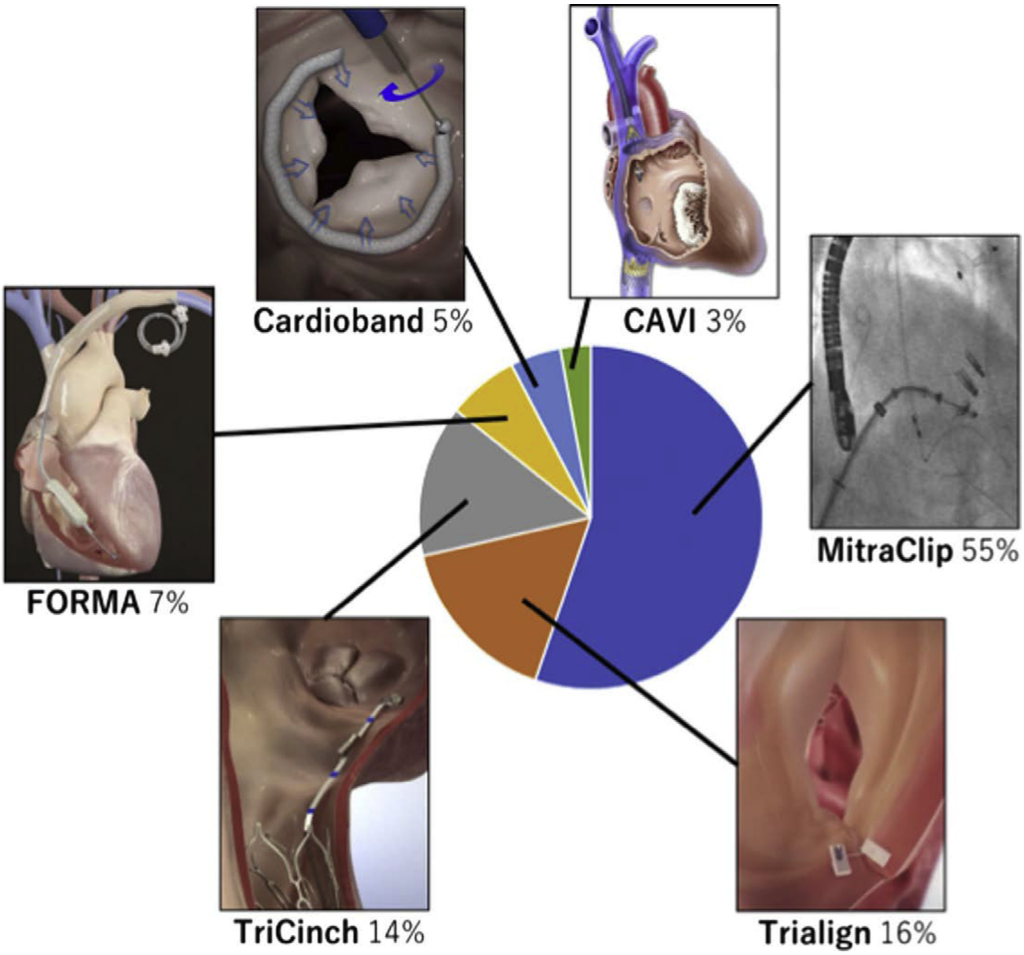

Figure 7. Currently investigated devices for interventional treatment of tricuspid valve regurgitation and their relative frequency of use in such procedures as collected in an international registry for interventional therapy of tricuspid regurgitation. Reproduced from Taramasso et al.[38] with permission from Elsevier.

was detected without evidence of persistent active endocarditis. Over the subsequent months a large pseudoaneurysm developed at the proximal graft anastomosis, with systolic compression of the ascending aorta (Fig. 8, C and E). The patient experienced increasing exercise intolerance. After careful interdisciplinary evaluation, it was decided to attempt device closure of the neck of the pseudoaneurysm because the risk of reoperation was expected to be very high. Given the cardiac anatomy with previous implantation of a biocomposite conduit, a transapical approach was chosen (Fig. 8, A and B). Deployment of 3 vascular plugs led to immediate complete occlusion of the pseudoaneurysm and improved exercise capacity (Fig. 8, D and F). This case vignette highlights that patients with congenital heart disease and previous complex surgical procedures may present at high risk for further surgery in case of recurrent complications. In such patients, careful interdisciplinary evaluation of treatment options and good collaboration between cardiac surgeons, interventionalists, and imagers are key to successful interventional procedures that may be much lower in risk than conventional surgery on cardiopulmonary bypass.

Outlook and Future Options

Pioneers in interventional cardiology and ingenious bioengineers are constantly developing novel techniques for percutaneous repair of failing heart valves. Many such devices have the potential to enhance our therapeutic armamentarium in the care of adults with congenital heart disease. These patients may particularly benefit from transcatheter treatment options because multiple prior open-heart surgeries put them at high risk for conventional open-heart redo surgeries. Key for success of such transcatheter procedures is the careful evaluation of the patient's individual anatomy and physiology and a multidisciplinary assessment involving adult congenital heart disease cardiologists, specialized imagers, cardiac surgeons, and interventionalists. Only this close collaboration will allow the optimal choice of the best treatment option in the individual patient. The high individual variability of cardiac anatomy, particularly within the native right ventricular outflow tract, poses a particular challenge and certainly requires the development of novel device designs and concepts.

Given that many interventions are relatively new, long-term outcomes are not yet well defined. This is of particular

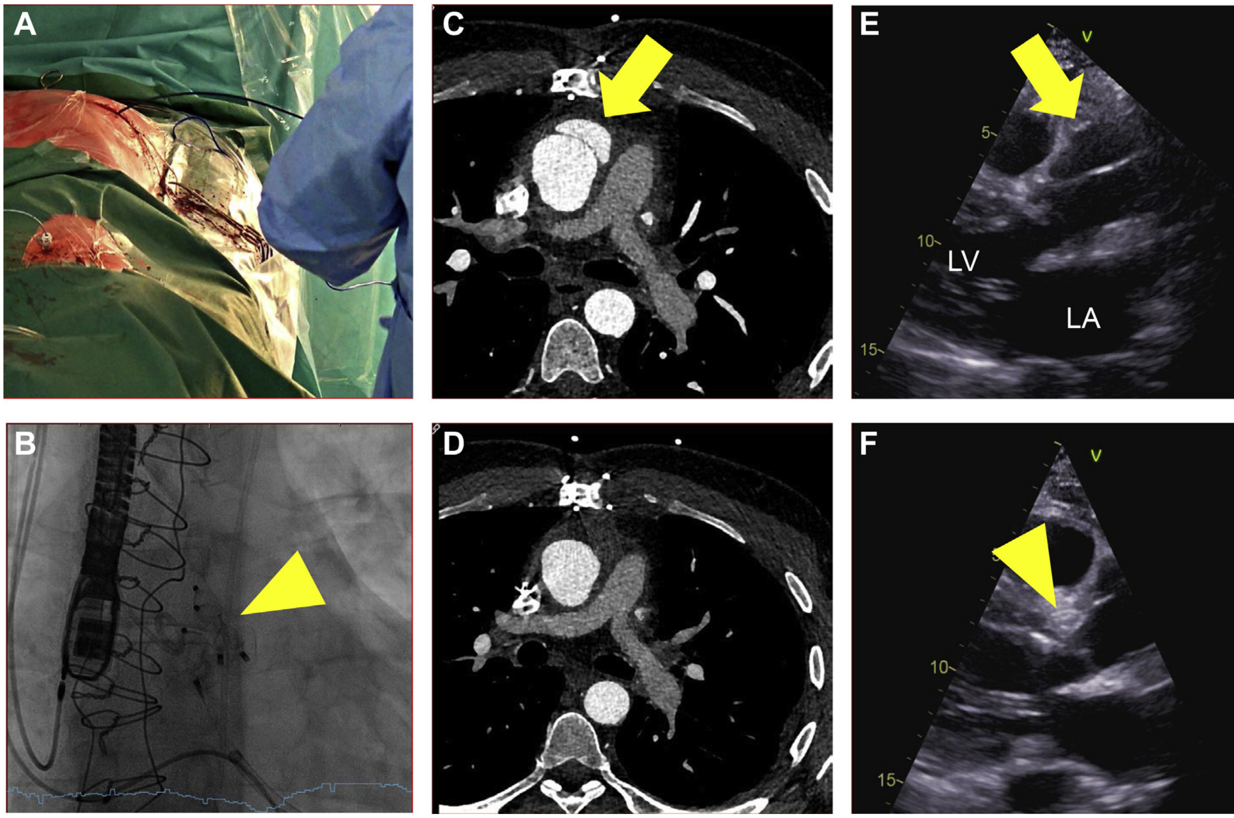

Figure 8. Transapical occlusion of a pseudoaneurysm after bioprosthetic composite graft replacement of the aortic root. (**A**) Transapical approach via minithoracotomy. (**B**) Fluoroscopy depicting deployment of the third vascular plug within the neck of the pseudoaneurysm at the proximal anastomosis of the composite graft (**arrowhead**). Computed tomography (**C**) before and (**D**) after percutaneous occlusion of the pseudoaneurysm (**arrow**). Parasternal views of transthoracic echocardiography (**E**) before and (**F**) after percutaneous occlusion of the pseudoaneurysm (**arrow** and **arrowhead**). Images **A** and **B** provided by courtesy of Francesco Maisano, Department of Cardiovascular Surgery, University Heart Centre, Zurich, Switzerland.

importance because many adults with congenital heart disease requiring valve interventions are relatively young and survival for many decades is expected. Therefore, careful and structured long-term observation of outcomes is important to refine the initial indications for a specific intervention and to potentially improve its technical aspects. Ever improving device technology and interventional skills with growing experience in these procedures will likely contribute as well to improved outcomes.

Disclosures

The authors have no conflicts of interest to disclose.

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