Aortic valve replacement in children: Options and outcomes

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Several disease pathologies such as congenital heart disease and rheumatic fever can affect the aortic valve (AV) in children frequently necessitating intervention. While percutaneous or surgical AV repair is recommended as initial management strategy in children with AV disease, AV replacement (AVR) might become necessary in children with significant valve destruction and after repair or intervention failure. AVR in children is associated with distinct clinical and technical problems owing to several anatomic, social and prosthesis-related issues.

In the current review, we list different AV substitutes, discuss their advantages and shortcomings, outline AVR results in children, and explore the divergence of outcomes in various age, anatomy and pathology subgroups; all in the aim to identify optimal AVR choice for each patient taking into consideration his unique anatomic and demographic characteristics.

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Keywords: Aortic valve replacement, Ross procedure, Rheumatic fever, Congenital heart disease, Mechanical valves

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Disclosure: Authors have nothing to disclose with regard to commercial support.

Received 2 April 2013; revised 19 August 2013; accepted 2 November 2013.
Available online 13 November 2013.
Introduction

The aortic valve (AV) can be commonly affected in children by several disease pathologies, frequently necessitating intervention [1,2]. The etiology of AV disease varies in different social and geographic locations; while congenital heart disease (CHD) is the most frequent pathology in Europe and North America, rheumatic fever (RF) remains a major pathology in developing countries [2]. Other less common pathologies include endocarditis, trauma and degenerative disorders.

Several developments in interventional cardiology allowed for early treatment of congenital aortic stenosis (AS), and recent experience with AV repair techniques demonstrated encouraging short-term and mid-term results in both congenital and rheumatic AV disease [3–6]. Current evidence supports AV repair as an initial palliative or lasting intervention in children with AV disease [3–6]. Nonetheless, in patients with significant valve destruction and after repair or intervention failure, AV replacement (AVR) is necessary [1,2]. AVR in children is associated with distinct clinical and technical problems owing to several anatomic, social and prosthesis-related issues.

In the current review, we list different AV substitutes, discuss their advantages and shortcomings, outline AVR results in children, and explore the divergence of outcomes in various age, anatomy and pathology subgroups; all in the aim to identify optimal AVR choice for each patient taking into consideration his unique anatomic and demographic characteristics.

Available valve substitutes

Optimal AV substitute in children would be one that’s readily available in different sizes, associated with excellent durable hemodynamic profile, has growth potential proportional to somatic enlargement, non-immunogenic, associated with minimal thrombo-embolism (TE) risk thus not requiring anticoagulation, and finally associated with excellent prosthesis longevity and low incidence of structural valve degeneration (SVD). No such choice is currently available to surgeons’ disposal and all valve alternatives are associated with important limitations.

Mechanical prostheses (MP)

MPs are readily available and come in different ranges with some small sizes (16 and 18 mm) suitable for young patients. Nonetheless, they’re still not suitable for infants and very small children who would require very small size prostheses that are not currently available at surgeons’ disposal. Their hemodynamic profiles vary with size and smaller prostheses have inferior flow properties. Special annular enlargement techniques could be utilized to allow placement of a larger prosthesis that could help with faster regression of left ventricular (LV) hypertrophy, improved LV function and improved symptoms. Those techniques include the Nicks procedure: aortic incision is extended to the area between the left/non coronary commissure and the base of the noncoronary cusp into the area of intervalvular fibrosa without cutting into the anterior mitral valve (MV) leaflet [7]; the Manougian procedure: almost same incision as in Nicks procedure but the cut is extended across the intervalvular fibrosa into the center of the anterior MV leaflet [8]; or the Konno procedure: the aortic annulus is incised between the right and left coronary cusps extending into the ventricular septum with patch reconstruction of the septum and ascending aorta [9].

Reported operative mortality of AVR with MP in children is 2–13% [1,10–14]; risk factors include younger age, presence of LV dysfunction and concomitant cardiac anomalies requiring surgery. Moreover, those children continue to have constant attrition rate with 15-year survival of 75–88% [1,10–14].

Most importantly, MPs require life-long anticoagulation to prevent TE. Anticoagulation in children can be challenging due to lack of compliance with medication and activity restraints therefore children could theoretically be at a higher risk of TE and anticoagulation-related bleeding complications. Nevertheless, despite those challenges, it seems that children are at lower risk of TE and bleeding than adults. In the literature, at follow-up ranging between 10 and 20 years, reported freedom from TE is 90–100% with average linearized rate of 0.3–0.7 per 100 patient-years and reported freedom from bleeding is 96% to 100% with average linearized rate of 0.3 per 100 patient-years [10–15]. It might be that children have different hemodynamic properties than adults with faster heart rate, less incidence of arrhythmias, atrial dilatation or myocardial dysfunction; all making them less prone to develop TE than adults. Those findings should be interpreted with caution given the fact that there is a continuous attrition risk in children who had received MPs with many late deaths sudden in nature, likely due to under-reported TE incidents.

Despite the lack of SVD, reoperations following AVR with MP in children are not uncommon...
and freedom from AV reoperation is 55–90% at 15 years [1,2,10–14]. Reoperations are usually related to development of patient-prosthesis mismatch (PPM) as the child grows older in the presence of fixed MP sizes that lack growth potential. Reoperations are also often indicated for pannus formation creating subvalvular obstruction, a complication that’s likely more common in children than in adults; in addition to few reported reoperations for valve thrombosis, paravalvular leak or endocarditis [1,2,10–14]. Nevertheless, considering other available AVR options, MPs continue to offer some of the highest reported freedom from reoperation rates.

**Tissue prostheses (TP)**

TPs are readily available and come in different ranges however they’re unavailable in sizes smaller than 19 mm, and hence are not suitable for small children even if annular enlargement techniques were utilized. Similar to MPs, their hemodynamic profiles vary with size and smaller prostheses have inferior flow properties.

TPs have a very low risk of TE and therefore do not require life-long anticoagulation. Nonetheless, their use in pediatric population is associated with decreased valve longevity and frequent need for reoperation due to lack of growth potential with development of PPM and most importantly SVD that’s faster than that seen in adults and inversely related to patient age and prosthesis size [2,4,16].

Alsoufi et al reported outcomes of 110 children who’ve undergone 123 AV or MV replacement with TP or homograft (HG). Among that patient cohort, 36 patients had AVR including 21 TP and 15 HG. Survival was 85% at 10 years while freedom from AV reoperation was about 35% at 10 years and 15% at 15 years with median TP longevity about 7.5 years. Overall 10-year freedom from TE and bleeding was 100% while freedom from endocarditis was 97%. The majority of patients were in New York Heart Association (NYHA) functional class I/II. Many females underwent successful uncomplicated pregnancies [16]. Those results suggested that while valve reoperation is inevitable, the lack of other optimal AV substitutes, favorable results such as low prosthesis-related morbidity rate, good long-term survival and functional status encourage consideration of TPs as valid replacement alternatives in selected children; especially teenage females and those with poor compliance with anticoagulation medications.

**Homografts (HG)**

HGs come in different ranges, are associated with excellent hemodynamic profiles that are similar for larger and smaller sizes and hence they’re suitable for the smallest children. Nonetheless, availability of HG, especially in the smallest ranges, varies due to the limited donor pool. They have a negligible TE risk and therefore do not require anticoagulation. Nonetheless, similar to TPs, they undergo rapid SVD leading to deterioration of their hemodynamic properties with development of obstruction and regurgitation, decreasing their longevity and requiring reoperation [2,4,16,17]. The fact that HG is resilient to infection make it the AVR of choice in patients having invasive endocarditis; however, HG use in pediatric population for other pathologies is very limited, especially that reoperations are challenging and associated with risks of bleeding and coronary injury. In the series from Riyadh, reoperation need for HG was similar to that of TP in children with only about 15% of patients free from AV reoperation at 10 years. Similarly, survival was about 83% and freedom from TE, bleeding or endocarditis was 100% [16].

**Ross procedure (Ross)**

Ross is an attractive AVR choice in children. It utilizes the pulmonary autograft (AG) in the aortic position and therefore it can be applied to all patients’ ages and is associated with excellent hemodynamic profile and cardiac recovery in all sizes. The growth potential of AG allows continuation of their superior hemodynamic profile long after Ross [18–21].

Furthermore, it’s a versatile operation that can be applied to patients with various left ventricular outflow tract (LVOT) and AV pathologies. The addition of a Konno-type aorto-ventriculoplasty in conjunction with Ross (Ross–Konno) allows successful management of patients with significant annular hypoplasia and those with complex multi-level LVOT obstruction [22–25]. The original Ross–Konno description involved a large incision in the “Konno” septum creating a ventricular septal defect (VSD) that was closed with a patch or anterior wall muscle extension of the harvested AG [22–25]. The modified Ross–Konno is currently utilized by many groups and involves an incision into the aortic annulus below the left/right commissure and extensive septal myectomy thus enlarging the LVOT without a VSD patch use [26].

The AG is implanted most commonly as a full root in children. Alternatively, the subcoronary or inclusion techniques could be utilized in older
children with adult-size aortic roots but not in young children with small aortic roots [27]. TE risk following Ross is negligible and therefore anticoagulation isn’t required [1,2,14,25,28–33].

Survival: Despite technical complexity, Ross has been shown to be safe in experienced hands. In the Ross registry and numerous contemporary series, reported operative mortality is under 2.5% [1,2,14,25,28–33]. Nonetheless, infants less than 1 year of age continue to have a high mortality risk approaching 15–20%, as compared to a mortality risk approaching 1% in children above 1 year of age [34–37].

Recent series demonstrated several risk factors for mortality in infants less than 1 year of age. Woods et al from Milwaukee reported Society of Thoracic Surgeons Congenital Heart Surgery Database outcomes of 160 infants who had undergone AVR including 145 Ross procedures (Ross–Konno in 101); overall hospital mortality was 16% and was 29% for neonates compared to 11% in older infants. Risk factors included concomitant arch surgery and post-operative ECMO [34]. Shinkawa et al from Ann Arbor reported 10-year survival of 77% and freedom from reoperation of 51% in 31 infants who had undergone Ross. Risk factors for death were concomitant MV and arch surgery [35]. Hickey et al from Toronto reported the Congenital Heart Surgeons Society (CHSS) results of 39 infants who had undergone Ross. They reported 5-year survival of 44% in neonates and 76% for infants older than 3 months. They noted several features that were associated with poor outcomes such as emergency surgery, coexisting MV pathology and LV dysfunction [36]. Alsoufi et al from Riyadh examined a similar patient cohort of 21 infants who had undergone Ross. Overall survival was 81% at 1 month and 63% at 5 years. Factors associated with poor outcomes included neonates, concomitant MV surgery, longer cross clamp time and post-operative ECMO. Of interest, Ross–Konno and prior surgical intervention weren’t associated with increased mortality risk. Freedom from reoperation was 86% at 5 years [37]. Those findings suggested that prior palliation with surgical or percutaneous aortic valvuloplasty should be attempted and might be associated with decreased mortality risk. They also suggested that outcomes following Ross in infants with complex LVOT obstruction could improve by better patient selection with expected long-term survival approaching 90% in those older than 3 months, isolated AV pathology not requiring concomitant MV or aortic arch surgery. Conversely, neonates with concomitant significant MV pathology or aortic arch obstruction might benefit better from other surgical alternatives such as the Yasui operation or even single ventricle palliation [37].

Time-related survival in children following Ross is stable with very little attrition risk beyond the perioperative period. Several series demonstrated that Ross in children offers the best long-term survival among other AVR substitutes [1,2,14,25,28–33]. This is mainly attributed to the excellent AG hemodynamic profile, trivial TE risk and lack of anticoagulation need.

Fate of the autograft: Despite excellent survival and superior hemodynamics following Ross, recommendation for this procedure has been declining, especially in older children in whom there are other AVR alternatives. This is mainly due to concerns about the development of neoaortic root dilatation, with or without subsequent AG regurgitation [18–21,32,38–44]. Until now it remains debatable whether the neoaortic annulus and root enlargement following Ross is actual growth in a manner proportional to somatic growth or pathological remodeling with disproportionate and excessive dilatation [18–21,43,44] it’s also debatable whether or not the pulmonary artery or aortic dimensions should be used as standard measurement reference points [18]. In general, neoaortic root dimensions immediately after Ross are larger than in healthy children, the annulus grows in proportion to somatic growth of the child while the root at the sinuses level and the sinotubular junction (STJ) dilates with time. In addition, AG regurgitation can develop in association with neoaortic dilatation, more so with STJ than sinuses dilatation [18–21,43,44]. The most remarkable demonstration of the fate of AG in children was reported by Pasquali et al from Philadelphia; they showed that at 6 years following Ross, freedom from neoaortic root Z score >4 was 3% only, freedom from AG regurgitation ≥ moderate was 60% and freedom from AG reintervention was 88% [38].

Several factors were identified to be associated with requirement for late AG reoperation. Those factors included STJ dilatation, bicuspid aortic valve with predominant aortic regurgitation (AI), dilated aortic annulus and geometric mismatch between semilunar valves (aortic larger than pulmonary) [23,32,38–45]. Moreover, older patients at time of Ross show faster increase in neoaortic root dimensions and aortic regurgitation [32]. It could be that AG might better adapt to the higher pressure in the aorta when it’s transitioned to the aortic position early at a younger age when smooth muscle cells are more prone to
differentiation and hyperplasia, thus allowing for the required structural changes in its wall to withstand aortic pressure. Moreover, children undergoing Ross at an early age often have a more severe multi-level LVOT obstruction with higher likelihood of having associated pulmonary hypertension. That pulmonary hypertension might have led to “priming” of the pulmonary root through structural changes in its wall thus allowing it to better adapt to the systemic blood pressure once it was transitioned into the aortic position. Finally, AG wall in older patients with higher systemic blood pressure might experience higher wall stress which could offer a stimulus for adverse remodeling and dilatation.

Children undergoing Ross–Konno comprise an interesting group of patients. By definition, those patients do not have an aortic annulus larger than the pulmonary valve, and most do not have primarily AI prior to Ross, thus they’re potentially at a lower risk of developing root dilatation and AG regurgitation. On the other hand, Ross–Konno involves incision across the annulus into the septum and placement of VSD patch with resultant loss of native annular support to AG and therefore potentially presents a higher risk of root dilatation and development of regurgitation. Fadel et al from Riyadh reported serial echocardiographic follow-up following modified Ross–Konno without VSD patch use; they demonstrated that both neoaortic annulus and root increase in size proportionately to somatic growth and that only few patients developed AG regurgitation that was usually mild and non-progressive with no requirement for AG reoperation [26].

Several surgeons adopted technical modifications aiming to reduce late AG dilatation when it’s implanted as a full root such as thinning of muscle rim below the valve, suturing AG within the native aortic annulus, shortening AG, enforcing proximal and distal suture lines with Dacron felt or replacing the ascending aorta with Dacron graft [27,32,46]. Reports in the literature suggested that AG stabilization and root inclusion or subcoronary implantation techniques might be associated with less incidence of AG dilatation and development of regurgitation [27]. Moreover, some surgeons suggested wrapping the AG with an absorbable mesh, glutaraldehyde-fixed bovine pericardium or encasing it in a Dacron tube to prevent dilatation [47]. Those techniques are suitable only for patients who won’t need AG growth because the outer shell prohibits this. Further follow-up is required to confirm the hypothetical advantages of those modifications.

In children with CHD, freedom from AG reoperation ranges between 75% and 95% at 10 years [1,2,14,25,28–33]. In a series by Alsoufi et al from Riyadh examining outcomes of 151 patients following Ross for CHD, 10-year survival was 92%, freedom from AG and right ventricle to pulmonary artery (RVPA) conduit reoperation was 95% and 71%, respectively [29]. In another series by the same group examining outcomes of 227 children following Ross for various etiologies, 10-year freedom from AG reoperation for patients with preoperative AS, AI and mixed disease was 97%, 69% and 93%, respectively. Risk factors for increased risk of AG reoperation were RF and earlier year of surgery. They demonstrated that AG longevity could be improved with better patient selection with expected freedom from reoperation in younger patients with predominant AS exceeding 95% compared to less than 50% in older patients with risk factors such as RF, AI and dilated aortic annulus [32].

Several series showed high failure and reoperation rates in children with RF undergoing Ross. In a recent study from Riyadh reporting outcomes of 104 children with RF who’ve undergone Ross; at 10 years, freedom from AG regurgitation ≥ moderate was 63%, freedom from AG reoperation was 65% for AI versus 90% for AS/mixed disease, freedom from RVPA conduit was 83% and from any cardiac reoperation was 53%. The majority of failures were evident in the first few years after Ross and the predominant pathology requiring AG reoperation was annular dilatation with cusp prolapse and failure of coaptation in 16/27, recurrence of inflammatory valvulopathy similar to that in RF in 8/27 reoperations. Patients with predominant preoperative AI, those with dilated annulus and concomitant MV regurgitation were at higher risk of HG failure. Those findings suggested that Ross should be used only selectively in children with RF with potential candidates being those with predominant AS, no dilated annulus, no associated MV disease and no active inflammatory markers [1,32,48].

Management of the failing AG depends on the mode of failure, status of the cusps and size of the neoaortic root. Patients with reasonably preserved AG cusps and regurgitation due to root dilatation and poor coaptation are candidates for AV sparing root replacement. In the past decade, there have been few series reporting successful reoperation on failing AG using the reimplantation or remodeling AV preservation techniques with good early outcomes [49,50]. Those techniques increase AG longevity despite the need
for aortic wall reintervention. Nonetheless, in patients in whom AG valve is irreparable due to recurrent inflammation, endocarditis, or extensive cusp thinning and destruction, autograft valve replacement (AVGR) is required. Choices for AVGR include redo root replacement (ARR) with a Bentall operation or AVGR with stented prosthesis (SP) that’s placed within the neoaortic root. Redo ARR is complex, associated with important risk of bleeding and coronary injury and could add significant risk, especially if concomitant cardiac surgery is needed at time of reoperation [51–53]. On the other hand, AVGR with SP placed within the neoaortic root is simpler and associated with lower risk of bleeding or coronary injury. Nevertheless, AVGR with SP would leave the neo-aortic root behind with concerns of continued dilatation, aneurysm formation and requirement for subsequent root reoperation. In a series from Riyadh examining the fate of the remaining neo-aortic root following AVGR with SP in 50 patients, there were no operative deaths and 10 years survival was 95%. Freedom from prosthesis, root and all-cause reoperations was 97%, 98% and 90% at 10 years, respectively. Serial echocardiographic data showed that there was little but progressive increase of remaining root and ascending aorta diameters however the requirement root reinterventions were very rare [54]. Those findings suggest that AVGR with SP is justified as it’s associated with low operative risk and good late outcomes and that redo ARR should be reserved with patients with significant root dilatation >4 cm at time of AG reoperation. Patients undergoing AVGR with SP should be followed for progressive root dilatation. Although published reports of AG dissection or rupture are rare, the incidence of those complications is likely to increase.

Fate of RVPA conduit: Reoperation for RVPA conduit change is a continuous problem following Ross. Longevity of RVPA conduit in patients following Ross is thought to be higher than that placed following repair of other congenital anomalies due to several factors such as the anatomic position of the conduit and the infrequent incidence of branch pulmonary stenosis. Several factors have been identified to be associated with RVPA conduit reoperation. Those included smaller conduit size, fresh homografts, aortic homografts and longer follow-up. In a meta-analysis by Takkenberg et al from Rotterdam, The average incidence of structural and non-structural degeneration of RVPA conduit in children was 1.6 (range 0.7–4.9) per 100 patient-years [31]. Reported freedom from RVPA conduit reoperation was 90–95% at 10 years and 75–85% at 15 years [1,2,14,25,28–33]. Decellularization of homografts was introduced in the early 2000’s and this process removed the endothelial cells and most of the other viable cells in the interstitial matrix without weakening the tissue. Initial experience suggests that those conduits have superior longevity compared to cryo-preserved ones and might further decrease reoperation rate.

In experienced hands RVPA conduit change is a relatively simple procedure and has been associated with low morbidity and operative mortality. RVPA conduit replacement will likely continue to be the most common type of reoperation following Ross. However, it’s important also to note recent advances in percutaneous pulmonary valve replacement that have allowed cardiologists to address this problem without surgical intervention. The immediate and short-term results of percutaneous pulmonary valve implantation are encouraging however longer follow-up is needed [55].

Cardiac reoperation: The initial thoughts were that reoperations following Ross would be limited to RVPA conduit change. Nonetheless, increased follow-up showed that reoperation was not as infrequent as anticipated and often involved multiple valves in addition to the coronaries and ascending aorta. Those reoperations could be specifically related to Ross, for example AG, RVPA conduit and coronary reoperations; or could be unrelated to Ross but rather to underlying cardiac pathology, for example MV and tricuspid valve reoperations. Ross is a complex operation and cardiac reoperations following Ross can be often more complicated [52,53,56]. A recent review from the Mayo clinic reported that 144 procedures were performed in 56 patients who required cardiac reoperations following Ross. Despite low hospital mortality (1 patient), surgical morbidity was high and results were beset by 4 additional deaths at a median follow-up of 8 months [52]. In a review from Riyadh, 50 patients underwent cardiac reoperations following Ross done in childhood. Risk factors for cardiac reoperation were RF, AI, concomitant cardiac surgery, use of fresh homografts and earlier era of surgery. Overall, 32 (55%) reoperations were isolated procedures whereas 26 (45%) were more complex involving 2–4 simultaneous cardiac procedures. In total, 92 procedures were performed including AVR (n = 31), RVPA conduit change (n = 23), MV replacement (n = 18), MV repair (n = 11), tricuspid valve repair (n = 5) and other (n = 4). There was 1 operative and 1 late death. Survival was 98% at 10 years. During
follow-up, 8/50 patients required further cardiac surgery following initial reoperation with freedom from additional cardiac surgery of 82% at 10 years [56]. That study demonstrated that despite complexity, Ross reoperation can be performed in experienced centers with low mortality and good mid-term results.

**Double valve replacement**

Several cardiac disorders, such as RF, endocarditis and CHD, can affect multiple valves and necessitate simultaneous double or triple valve surgery. Double valve surgery is challenging, often complicated and prolonged due to previous surgeries and common requirement for concomitant procedures such as reconstruction of valve annuli or the base of the heart secondary to infection, calcification, fibrosis, or insufficient space to secure the placement of prostheses of adequate size. Moreover, early post-operative care of those patients is often difficult due to the frequent presence of poor preoperative clinical and hemodynamic status that might further imperil outcome.

Only few series in the literature describe outcomes following simultaneous AV and MV replacement in children. In a recent study from Riyadh reporting outcomes following double valve replacement in 84 children; operative mortality was 4%, 15-year survival was 78% (92% for TP and 76% for MP) while 15-year freedom from reoperation was 59% (70% for MP and 0% for TP). Freedom rates from endocarditis, TE and bleeding complications at 15 years were 90%, 92%, and 96%, respectively. Among survivors, 95% were in NYHA class I/II. Those findings indicated that while reoperation risk was higher, freedom from other valve related complications was similar to that in patients who’ve undergone single valve replacement of the same type [57].

**Comparisons of different aortic valve alternatives**

Few studies in the literature compared outcomes of various AVR options in children. Karamlou et al from Toronto reported results following AVR with several alternatives in 160 patients; on multi-variable analysis, Ross was associated with improved survival while HG or TP use was associated with increased reoperation [2]. Ruzmetov et al. from Indianapolis compared outcomes following AVR with several alternatives in 147 patients; they found that 10-year survival was highest following Ross (98%) as compared to MP (88%), HG (87%) or TP (82%) while freedom from reoperation was highest following Ross (91%) as compared to MP (88%), HG (87%) and TP (40%) [14]. Alsoufi et al from Riyadh compared outcomes in 346 children following AVR (215 Ross and 131 MP). After propensity adjustment, MP use was a risk factor for early and late death while Ross was a risk factor for reoperation; however they had a large proportion of children with RF to account for the increased reoperation rate after Ross [1]. Lupinetti et al from Seattle compared outcomes following 51 AVR in children (26 MP, 19 Ross, 6 HG); they found lower 2-year freedom from reoperation in MP group (80%) as compared to Ross and HG (96%) [58]. In another study, the same group compared outcomes between children undergoing AVR with Ross (n = 78) or HG (n = 25); they found equal 7-year survival for both groups (96%) but superior freedom from reoperation for Ross (88%) vs. HG (73%); and also demonstrated that Ross was associated with superior maintainable hemodynamics over HG that showed deterioration of hemodynamic profile in conjunction with SVD [17].

**Summary and recommendations**

In summary, AVR in children creates a lifelong disease that continues throughout adulthood. Each AV substitute has recognized shortcomings that affect late survival and reoperation need. The choice of AV substitute should be carefully deliberated at time of first AV intervention taking into consideration each patient’s unique anatomic, clinical and social characteristics, and acknowledging the fact that many selected management plans would require eminent reoperation necessitating lifetime observation.

In children with AV disease requiring operation, AV repair should be attempted as a temporizing or lasting initial treatment as it improves symptoms, stabilizes the heart and allows delay in AVR need till an older age when surgery is associated with lower risk and different replacement options can be offered. AVR is necessary in patients in whom AV could not be repaired or those following repair failure. Neonates, infants and small children with AS requiring AVR should undergo Ross or Ross–Konno. Although Ross is associated with clear survival advantage over other AVR alternatives in that patient population, operative mortality for Ross is the highest in that subgroup of patients, especially if concomitant MV or arch surgery is required. Selected neonates and small infants with significant coexisting MV and arch anomalies should be considered for alternative management strategies that could include single ventricle palliation. In older
children, Ross continues to offer survival and reoperation advantage over other alternatives in patients with AS or mixed disease after a prior palliation. Nonetheless, AVR with MP might be a superior choice in patients with predominant AI and dilated AV annulus. AVR with TP could also be considered in that setting especially in females and children incompetent with anticoagulation regimen. HG use is reserved for infants and small children in whom the pulmonary AG could not be used or in cases with invasive endocarditis. In children with RF, AVR with MP or TP should be contemplated while Ross should be offered only in a selected group of patients with no active inflammation, no AI, no dilated AV annulus, and no concomitant involvement of other cardiac valves.

References


