REVIEW

Valve replacement in children: A challenge for a whole life

Remplacement valvulaire chez l’enfant : les défis pour toute une vie

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Summary Valvular pathology in infants and children poses numerous challenges to the paediatric cardiac surgeon. Without question, valvular repair is the goal of intervention because restoration of valvular anatomy and physiology using native tissue allows for growth and a potentially better long-term outcome. When reconstruction fails or is not feasible, valve replacement becomes inevitable. Which valve for which position is controversial. Homograft and bioprosthetic valves achieve superior haemodynamic results initially but at the cost of accelerated degeneration. Small patient size and the risk of thromboembolism limit the usefulness of mechanical valves, and somatic outgrowth is an universal problem with all available prostheses. The goal of this article is to address valve replacement options for all four valve positions within the paediatric population. We review current literature and our practice to support our preferences. To summarize, a multitude of opinions and surgical experiences exist. Today, the valve choices that seem without controversy are bioprosthetic replacement of the tricuspid valve and Ross or Ross-Konno procedures when necessary for the aortic valve. On the other hand, bioprostheses may be implanted when annular pulmonary diameter is adequate; if not or in case of right ventricular outflow tract discontinuity, it is better to use a pulmonary homograft with the Ross procedure. Otherwise, a valved conduit. Mitral valve replacement remains the most problematic; the mechanical prostheses must be placed in the annular position,

Abbreviations: AV, auriculoventricular; AVR, aortic valve replacement; LVOT, left ventricular outflow tract; MV, mechanical valve; MVR, mitral valve replacement; PA, pulmonary autograft; PH, pulmonary homograft; PVR, pulmonary valve replacement; RVOT, right ventricular outflow tract; TV, tricuspid valve; TVR, tricuspid valve replacement; VR, valve replacement.

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avoiding oversizing. Future advances with tissue-engineered heart valves for all positions and new anticoagulants may change the landscape for valve replacement in the paediatric population.

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**Background**

Valvular pathology in infants and children poses numerous challenges to the paediatric cardiac surgeon. Without question, valvular repair is the goal of intervention because restoration of valvular anatomy and physiology using native tissue allows for growth and a potentially better long-term outcome. When reconstruction fails or is not feasible, valve replacement (VR) becomes inevitable. Which valve for which position is controversial. Homograft and bioprosthetic valves achieve superior haemodynamic results initially but at the cost of accelerated degeneration. Small patient size and the risk of thromboembolism limit the usefulness of mechanical valves (MVs), and somatic outgrowth is a universal problem with all available prostheses. The goal of this article is to address VR options for all four valve positions within the paediatric population. We review current literature to support our preferences.

**Aortic valve replacement**

Aortic valve disease is one of the most common congenital cardiac defects, occurring in 5% of all children with heart disease. The bicuspid aortic valve is the second most common pathological valve entity in the paediatric patient population that requires VR in a high percentage of patients over their lifetimes [1].

**Ross procedure**

Controversy over prosthetic type for aortic valve replacement (AVR) has dropped dramatically in the past 15 years because of the growth in popularity and excellent results obtained with the Ross procedure (Fig. 1A and B). Pulmonary autograft (PA) has become the first choice of AVR in children and adolescents in some institutes [2,3]. PA shows excellent haemodynamic performance, superior longevity (Fig. 2) [4], freedom from anticoagulation and haemolysis and decreased susceptibility to endocarditis. PA is also known to have the potential for growth. However, the Ross procedure is a technically demanding procedure and reoperation for bleeding and postoperative conduction abnormality is not as rare as early complications. Freedom from autograft dysfunction, including severe autograft insufficiency, ranges from 75% to 100% depending upon the duration of follow-up [5]. Elkins et al. [6] reported freedom from autograft replacement of 93% and freedom from severe autograft insufficiency or valve-related death of 90% at their 12-year follow-up. Autograft insufficiency is one of the leading causes of reoperation with the Ross procedure and several factors are implicated as risk factors, such as preoperative diagnosis of aortic insufficiency, presence of dilated aortic annulus, bicuspid aortic valve, rheumatic heart disease, technical imprecision, the type of insertion and inherent disease of the pulmonary valve. Elkins et al. reported a freedom from right ventricular outflow tract
Figure 1. Ross intervention. After harvesting the pulmonary autograft and left and right coronary arteries (A), the left ventricular outflow tract is reconstructed with the pulmonary autograft and reimplantation of the coronary arteries, and the right ventricular outflow tract is reconstructed with a pulmonary homograft (B). By courtesy of Hodder Education, London.
reinforcement with homografts (L VOT) provides normalization of the aortic valve. Means ± 67% confidence limits for three time intervals are indicated [19].

Figure 3. Kaplan-Meier graph depicting freedom from all valve-related reoperations after homograft or autograft replacement of the aortic valve.

Bigger than that of in situ insertion. The Konno procedure requires incision of the ventricular septum, which might cause ventricular dysfunction or conduction abnormality. In the Manouguian procedure, the incision is extended to the anterior mitral leaflet and might cause mitral insufficiency. The Yamaguchi procedure does not damage either the ventricular septum or the mitral leaflet.

Shanmugam et al. [11] reported that no rereplacement of prosthesis was required when the patient received a prosthestesis 21 mm or larger in size. Masuda et al. [15] reported that freedom from rereplacement of aortic valve was 94% at 15 years and was at least compatible with the results of other series with mechanical prostheses by Shanmugam et al. (92% at 20 years) [11] and Ruzmetov et al. (84% at 19 years) [16], and was not inferior to the results of PA reported by Elkins et al. (93% at 12 years) [6] and Pasquali et al. (81% at 8 years) [7]. An actuarial survival rate of 92% and a freedom from valve-related complications rate of 86% at 15 years seem quite acceptable [15]. Regression of left ventricular dilatation in children with severe aortic regurgitation can be observed on echocardiography and magnetic resonance imaging after timely AVR [17].

Concerning anticoagulation problems, Akhtar et al. [18] reported a study assessing long-term survival and anticoagulant-related complications after mechanical VR in adolescents with rheumatic heart disease. Patient survival rates at 30 days, 3 months and 1, 5 and 10 years were 95.5%, 93.2%, 87.5%, 82.9% and 82.9%, respectively. MV thrombosis occurred in 4.5% patients and was fatal in 3.4% of them. Severe haemorrhage required hospital admission in 4.5% of patients.

Although quite durable, MVs require chronic anticoagulation, which can be poorly tolerated and quite difficult to control in some children. Patient growth and acquired patient-prosthesis mismatch are not uncommon problems with mechanical AVR.

Bioprostheses, homografts and xenografts

Bioprostheses, homografts and xenografts in children and adolescents have been largely abandoned due to accelerated degeneration (Fig. 3) [19]. In the younger age group,

Mechanical aortic valve replacement

MVAs are reserved for children who have connective tissue disorders or whose native pulmonary valves are unsuitable for translocation to the aortic position. AVR using mechanical prosthetic valves in children often requires annular enlargement to insert commercially available prostheses [11]. The Yamaguchi [12], Manouguian [13] and Konno [14] procedures enable insertion of prostheses two sizes

(RVOT) homograft replacement of 90% at 12 years for children. Rates of freedom from RVOT were also similar for other authors [7]. Because of the diminishing availability of homografts, several conduits are used to reconstruct RVOT; however, their durability seems to be worse than that of homografts [5]. Finally, Elkins et al. [6] reported freedom from all valve-related morbidity of 79% at 11 years.

Husain et al. [8] reported that freedom from replacement of the PA was 96% at 10 years. Freedom from replacement of the pulmonary homograft (PH) was 96% at 10 years.

Technical modifications, such as resection and graft replacement of a dilated ascending aorta, annular reinforcement with circumferential felt or Dacron and/or reinforcement of the entire autograft root, are all options to minimize autograft dilation and insufficiency.

In neonates and infants with left ventricular outflow tract (LVOT) obstruction, the Ross procedure, although more complex, provides excellent normalization of haemodynamics and regression of left ventricular hypertrophy by avoiding residual lesions [9]. Despite the need for reoperation and potential for autograft root dilation, the Ross and Ross-Konno procedures remain the best choice for AVR in infants with multilevel LVOT obstruction or severe aortic insufficiency following valvuloplasty [10].
there is a significant risk of structural valve deterioration, reported to range from 71% to 87% at 10 years [20].

Decellularized aortic valve allografts [21] appeared to be more resistant to calcification and did not show any major structural and morphological alteration. If these results are confirmed with longer follow-up periods, this technique may be a promising alternative to AVR for a selected group of patients, especially females [22].

Mortality

Karamlou et al. [4] identified that younger age and lower weight at initial AVR unfavourably influenced mortality without repeated replacement, especially in the extreme cases of very young age or very low weight. Previously published reports, which showed that neonates and those aged less than 6 months compose the highest-risk group, agree with these findings [1]. There are several reasons for poor outcome in this population. First, young age at initial operation was significantly associated with the presence of other cardiac anomalies, including important mitral valve dysfunction, which accounted for substantial mortality. Others have noted that those with concomitant cardiac lesions fare worse than those with isolated aortic valve disease [1]. Second, the preoperative clinical status of younger patients, especially neonates, is likely to be considerably worse than those undergoing later AVR. Correlation between poor preoperative left ventricular function (fractional shortening < 25%) and late mortality has been established. Finally, younger patients (and those with lower weight) are at highest risk of prosthesis outgrowth necessitating subsequent repeated replacement or intervention, which may contribute to increased mortality. The need for concomitant aortic arch reconstruction or augmentation was also identified as an incremental risk factor for death without a second AVR.

In a recent study, Alsoufi et al. [23] reported on 346 children who underwent AVR (215 Ross procedures; 131 placements of a mechanical prosthesis). Patients undergoing the Ross procedure were younger, more likely to have a congenital cause and less likely to have a rheumatic or connective tissue cause; they had a lower frequency of regurgitation, required more annular enlargement and had less concomitant cardiac surgery. Competing-risk analysis showed that 16 years after AVR, 20% of patients had died without subsequent AVR, 25% had undergone a second AVR and 55% remained alive without further replacement. Factors associated with early-phase death included MV and a non-rheumatic cause. MVs were also associated with constant-phase mortality. Repeated AVR was associated with the Ross procedure and a rheumatic cause. In children who received a mechanical prosthesis, younger age and smaller valve size were significant risk factors for death. Freedom from homograft replacement after the Ross procedure was 82% at 16 years of follow-up. Results from this study showed good outcomes and an acceptable complication rate with both valve choices. Given the significantly increased risk of early and late death in younger children receiving smaller MVs, the Ross procedure confers a survival advantage in this age group at the expense of increased reoperation risk, especially in patients with a rheumatic cause.

Mitral valve replacement

Different congenital malformations may affect the mitral valve either in isolation or in association with other cardiac anomalies [24]. Improvements in surgical techniques have made it possible to obtain good results when a mitral repair is required. Anatomical analysis is of particular importance for surgical management and prognosis. As a result, the need for mitral valve replacement (MVR) is relatively uncommon in children. But in some cases, MVR is the last recourse. The most common indications for MVR in children include rheumatic disease, endocarditis, mitral stenosis in Shone’s syndrome or failed aortic-ventricular (AV) canal repair. MVR carries the highest mortality for any paediatric AVR and has a much poorer long-term prognosis than any other VR in children. The reported operative mortality for MVR in infants is 5% to 52%. The 5- and 10-year survival for these patients has been reported as 33% to 95% (Table 1) [25–36]. Because of these concerning statistics, alternatives to MVR should include aggressive attempts at valve repair and sometimes conversion from biventricular to single ventricle repair or even cardiac transplantation [30]. MVR is common in small children who have a small mitral annulus. Unfortunately, annular enlargement options are sparse. Attempting to oversize the prosthesis at the time of MVR can produce subaortic obstruction and should be avoided. Prosthetic leaflet entrapment and conduction block after MVR pose significant postoperative morbidity and mortality. Common reasons for reoperation include prosthetic stenosis, thrombosis and endocarditis. In comparison with initial MVR, the mitral annulus can usually be upsized 2 to 3 mm in diameter at the time of redo-MVR. Low-profile bileaflet pyrolytic carbon valves are the most popular prostheses for MVR; however, all MVs require lifelong anticoagulation. Bio-prosthetic xenografts and mitral homograft valves do not require anticoagulation but have limited durability of 3 to 5 years in the mitral position [29].

In an attempt to find a more durable tissue valve that does not require lifelong anticoagulation, some centres used the PA MVR or Ross II technique [8] in selected older children and young adults whose pulmonary valves are large enough for MVR (20 mm in diameter). Ross II does not require anticoagulation and has other potential advantages, such as providing a flexible prosthesis, thus removing the need for a rigid structure in the mitral annulus. However, experience is scarce.

Survival

The largest review of MVR in the young (< 5 years of age) paediatric population was published by Caldarone et al. [30], who analysed data gathered by the Pediatric Cardiac Care Consortium (45 centres; 1982–1999). MVR was performed 176 times in 139 patients, all less than 5 years of age. Operative morbidity in these patients included heart block requiring pacemaker implantation (16%), endocarditis (6%), thrombosis (6%) and stroke (2%). The diagnosis of complete atrioventricular septal defect, Shone’s syndrome, an increased ratio of prosthetic size/weight and supra-annular position were all found to be statistically significant predictors of early mortality.
Table 1  Literature review of postoperative mortality and long-term survival after mitral valve replacement.

<table>
<thead>
<tr>
<th>Article</th>
<th>Number of patients</th>
<th>Postoperative mortality (%)</th>
<th>5-year survival (%)</th>
<th>10-year survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Younger</td>
<td>Older</td>
<td>Younger</td>
<td>Older</td>
</tr>
<tr>
<td>Caldarone et al., 2001 [30]</td>
<td>&lt; 5 years (n = 139)</td>
<td>18</td>
<td>75</td>
<td>74</td>
</tr>
<tr>
<td>Alexiou et al., 2001 [26]</td>
<td>&lt; 5 years (n = 23)</td>
<td>5 – 16 years (n = 21)</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Erez et al., 2003 [31]</td>
<td>&lt; 2 years (n = 29)</td>
<td>2 – 18 years (n = 61)</td>
<td>52</td>
<td>3</td>
</tr>
<tr>
<td>Vohra et al., 2006 [36]</td>
<td>&lt; 5 years (n = 24)</td>
<td>21</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Beierlein et al., 2007 [29]</td>
<td>&lt; 2 years (n = 21)</td>
<td>2 – 18 years (n = 23)</td>
<td>41</td>
<td>6</td>
</tr>
<tr>
<td>Ackermann et al., 2007 [29]</td>
<td>&lt; 6 years (n = 69)</td>
<td>11</td>
<td>65</td>
<td>65</td>
</tr>
<tr>
<td>Selamet Tierney et al., 2008 [35]</td>
<td>&lt; 5 years (n = 118)</td>
<td>75</td>
<td>56</td>
<td></td>
</tr>
<tr>
<td>Alsoufi et al., 2010 [28]</td>
<td>&lt; 2 years (n = 37)</td>
<td>2 – 8 years (n = 42)</td>
<td>30</td>
<td>6</td>
</tr>
<tr>
<td>Henaine et al., 2010 [32]</td>
<td>&lt; 5 years (n = 29)</td>
<td>13</td>
<td>86</td>
<td>86</td>
</tr>
<tr>
<td>Rafii et al., 2011 [34]</td>
<td>&lt; 2 years (n = 18)</td>
<td>2 – 18 years (n = 27)</td>
<td>11</td>
<td>0</td>
</tr>
<tr>
<td>Alsoufi et al., 2011 [27]</td>
<td>&lt; 2 years (n = 36)</td>
<td>2 – 18 years (n = 271)</td>
<td>26</td>
<td>4</td>
</tr>
<tr>
<td>Kanter et al., 2011 [33]</td>
<td>&lt; 4 years (n = 15)</td>
<td>7</td>
<td>87</td>
<td></td>
</tr>
</tbody>
</table>

In our centre [32], the comprehensive mortality is 17%. Among five deceased children, two had a complete AV canal and one had a partial AV canal; one child had Shone’s syndrome with several previous operations, among them a Ross procedure for the left outflow tract (the delayed treatment resulted in a major alteration of the function of the left ventricle); and the last child had a hammock mitral valve associated with an aortic valvular stenosis. Therefore, mortality is linked to the complexity of associated cardiopathies [37].

On the other hand, Rafii et al. [34] found that there was no significant difference in survival between patients aged less than 2 years and patients aged 2 to 18 years; age less than 2 years remains a risk factor for reoperation but not for mortality.

Conversely, Alsoufi et al. (Fig. 4) [27] in a recent study of 307 children who underwent MVR, showed that factors for mortality were: younger age (< 3 years) at time of MVR; longer cross-clamp time; postoperative complications (complete heart block, bleeding and low cardiac output); and higher prosthesis size/body surface area—predicted mitral annulus size. Of importance, patients with underlying congenital mitral valve disease had worse survival compared with those with other pathologies, especially rheumatic fever.

Ackerman et al. [25] found that age, weight, body surface area, predicted annulus diameter, prior surgery, underlying disease and ratio of prosthetic valve diameter to body weight were significant predictors of death. Variables associated with rereplacement of the systemic atriioventricular valve were body surface area, prosthetic valve diameter, predicted annulus diameter and presence of multiple left-sided obstructive lesions. The majority of patients received a prosthesis that was larger than

Figure 4.  Competing-risks depiction of outcomes following mitral valve replacement (MVR) in children aged 1 to 18 years. The competing risks for the two events (mitral reoperation and death without mitral reoperation) showed that at 10 years following MVR, approximately 15% of patients had died and 24% had undergone mitral reoperation, while at 20 years following MVR, approximately 17% of patients had died, 51% had undergone mitral reoperation and only 33% were alive and free from mitral reoperation [27]. MV: mitral valve.
the predicted annulus diameter. There was good correlation between the prosthetic valve diameter and the measured annulus diameter. Mismatch, as described by the difference in z scores of prosthetic valve diameter and measured annulus diameter, was not a significant predictor of death or rereplacement of the systemic atrioventricular valve.

**Annular or supra-annular**

Supra-annular MVR is useful for children with a small annulus. Kanter et al. [33] found that operative survival was good with infrequent heart block. Complications are common, including pulmonary vein stenosis and the need for LVOT obstruction relief. Pulmonary vein stenosis is a marker for poor outcome; all patients without pulmonary vein stenosis survive long term. Selamet Tierney et al. [35] showed that supra-annular MVR was associated with worse survival than annular MVR. Supra-annular placement should be reserved only for those patients in whom the native annulus is too small to accept a commercially available valve.

**Anticoagulants**

The administration of anticoagulants (antivitamin K type) after MVR is essential. Drawbacks and potential complications related to anticoagulants should be considered, but literature [26,32] and experience in the use of anticoagulants in MV, associated with rigorous medical observance and surveillance, show that anticoagulants in the paediatric age group are tolerated.

**Pacemaker placement**

Morphological investigation [38] showed that the AV node artery runs in close proximity to the annulus in 23% of cases. Damage to the AV node artery may play a role in the development of AV block. In our study [32], four children (13%) had a complete AV block requiring the placement of a permanent pacemaker during the first intervention. Selamet Tierney et al. [35] showed that patients with a supra-annular prosthesis had a lower risk of pacemaker placement in the early post-MVR period than those with an annular prosthesis, but remained at risk when the prosthesis was subsequently replaced (Fig. 5) [35].

**Redo-MVR**

We performed a reintervention for MVR in 10 patients (37%); among them, eight patients (27%) have been reoperated on due to the MV becoming restrictive. All these patients had a new MV at least two sizes larger than the initial valve. No mortality was observed during the reoperation of patients. We showed that the mitral ring may expand during growth even if there is a prosthesis all around, which allowed the placement of a bigger prosthesis during the ulceration operation. Raghunee et al. have confirmed this point [39]. However, precautions must be taken not to use oversized prostheses to avoid circumflex compression.

In a multi-institutional study [30,39], the potential univariate predictors of second MVR were: younger age; lower weight; increased prosthetic valve size/body weight ratio; Shone’s syndrome; smaller prosthesis; and prosthesis other than St. Jude’s. In our study [32], we found that at first intervention, weight less or equal to 7 kg and age less or equal to 1 year were associated with redo-MVR. Our low level of redo-MVR (76.7% at 15 years) compared with other studies [29,35] may be explained by our strategy of waiting as long as possible because the transmitral gradient increases very slowly and keeps patients asymptomatic for a long period of time.

**Right ventricular outflow tract reconstruction and/or pulmonary valve replacement**

The pulmonary valve is the most common valve replaced in the congenital population. Pulmonary valve replacement (PVR) continues to be a ‘weak link’ in our management of children and adults with congenital heart disease. PVR for a newborn with truncus arteriosus is vastly different from PVR for an adult undergoing reoperation after tetralogy of Fallot repair in childhood.

A PH was the PVR option of choice for congenital PVR from the mid-1980s to 2000. Although early homograft regurgitation in non-Ross AVRs was a common finding, regurgitation was rarely the cause of valve failure or an indication for PVR. Obstruction was the most common indication for PVR after using a PH. Most large series report the need for rereplacement of a PH in non-Ross patients within 5 to 7 years. This, however, is not the case for PHs implanted during a Ross AVR because the PH can be oversized and placed in a true orthotopic position.

Evaluating the durability of the PH has been critical in determining the choice of prosthesis for PVR. Forbes et al. [40] reviewed 185 consecutive PH implants at a single
institution over 14 years. Three separate age groups were evaluated. Their analysis showed that smaller homograft size, younger aged patients and the diagnosis of truncus arteriosus were all risk factors for homograft failure in a univariate analysis. Smaller homograft size was the only predictor of failure in multivariate analysis. Ross procedure-related implants had the best outcomes, with a 5-year PH survival of 94% (Fig. 6A). Non-Ross PH implants in children older than 10 years had a graft survival of 76% (Fig. 6B).

The largest published series of PVR and right ventricle-to-pulmonary artery conduits came from Dearani et al. [41] at the Mayo Clinic, who described late follow-up for 1095 patients undergoing right ventricle-to-pulmonary artery conduit or PVR over a period of 37 years. During this period, 1270 RVOT procedures were performed.

In the most recent decade, mortality was 3.7%. Mean follow-up for the entire series was 10.9 years, with a maximum of 29 years. Three types of conduits were used: 730 patients received porcine valved Dacron conduits, 239 received PHs and 126 had non-valved conduits implanted. Risk factors cited for conduit failure included use of a PH, younger age at initial operation and smaller conduit size. An additional RVOT reconstruction option often used by Husain and colleagues [8] is the insertion of a Gore-Tex monocusp. Transannular incisions were required in 40% of children with tetralogy of Fallot and its variants. The authors found that use of the Gore-Tex monocusp minimizes early and midterm valvular regurgitation, as well as allowing for right ventricular functional recovery. Freedom from reoperation has ranged from 87% to 92% at 10 years based on subgroup analysis of RVOT pathology.

The Contegra bovine right ventricular pulmonary artery conduit is a glutaraldehyde-preserved bovine jugular venous-valved conduit. Size availability ranges from 12 to 22 mm in diameter. It is very 'user friendly', there is no requirement for postoperative anticoagulation and the valve retains better competence than a PH. Initial results from Brown et al. [42] have been quite promising; their Contegra series included 85 implants in patients ranging in age from 2 weeks to 18 years. There have been two early (3%) and four late (7%) deaths, with none being conduit-related. There has been only one Contegra conduit explantation for a right ventricular pseudoaneurysm. Seven patients have required reinterventions to relieve stenosis at or beyond the distal conduit anastomosis; six of these seven patients required balloon dilatation for branch level stenosis beyond the conduit itself. Thus far, they have not seen any evidence of conduit shrinkage or significant dilation.

**Hancock porcine valved conduit**

Belli et al. [43] have shown the long-term outcome of 214 patients with the Hancock porcine valved conduit. Median age at operation was 62.5 months (range, 1 week to 50 years), including 14 neonates (6%). The higher RVOT systolic pressure gradient at discharge did not influence conduit longevity. Conduit reoperation was delayed due to percutaneous balloon dilatation in 14 patients, associated with stenting in seven patients. Survival with freedom from conduit reoperation was 98% at 1 year, 81% at 5 years and 32% at 10 years.

During the study period, the Hancock conduit was explanted and replaced in 88 patients at a median delay of 72.1 months (range, 5 to 142 months) (Fig. 7).

The Hancock valved conduit is a safe and reliable alternative to homografts; it appears to be appropriate in patients with a limited pulmonary vascular bed and high pulmonary artery pressures. Caution is required in neonates because of the rigidity of the Dacron housing. Initial results with secondary percutaneous procedures are encouraging. Other advantages of the Dacron housing are that it allows a safer approach and easy removal during conduit reoperation and the limitation of the late conduit calcification was only at the valvular level. The authors recommend its use particularly beyond the neonatal period, in the presence of a restrictive pulmonary vascular bed, increased pulmonary vascular resistance or both and, finally, as a palliative right ventricular pulmonary artery conduit.
Valve replacement in children

Mechanical valves

MVs and valved conduits have been used in the pulmonary position for highly selected older children and adults. Chronic anticoagulation requirements make this option less attractive. Haas et al. [44] published a series of 15 patients over a 5-year period who received a MV conduit in the pulmonary position. All patients had a mean of 3.0 ± 1.2 previous operations. All patients survived the operation. At follow-up of 11 to 63 months, all but two patients had normal right ventricular function, with a mean gradient of 14 ± 9 mmHg across the pulmonary MV. All patients were on anticoagulation therapy with a goal international normalized ratio of 3.0 to 4.5. Overall, this option seems feasible for highly selected patients, especially those somewhat older patients with multiple previous operations, patients who have other mechanical heart valves and patients with social and family support systems to monitor and administer anticoagulation therapy.

Bioprostheses

Surgical relief of RVOT obstruction in tetralogy of Fallot often includes a transannular incision extending from the muscular infundibulum to the main pulmonary artery. Although the afterload to the right ventricle is reduced, the tradeoff is pulmonary valvar incompetence, long-term pulmonary regurgitation and progressive right ventricle dilation. Right ventricle dilatation is associated with vulnerability to arrhythmia; repaired tetralogy of Fallot patients are known to be at increased long-term risk of mortality. A potential solution to this problem is PVR, with the goal of reversing the process of right ventricle dilation. In a matched comparison with a similar tetralogy of Fallot group, late PVR for symptomatic pulmonary regurgitation/right ventricle dilation did not reduce the incidence of ventricular tachyarrhythmia or death [45].

Our main policy is to perform PVR with a stented bioprosthesis (Mitroflow® or Triflecta®) with pulmonary branch plasty when required and with direct closure or with haemashield patch of the RVOT; this PVR is made by sternotomy except for special cases [46], with extracorporeal circulation and without cross-clamping of the aorta. The timing of the intervention should be as late as possible. Thus, between 2001 and 2011, 60 patients in our institution underwent a PVR after the repair of tetralogy of Fallot in infancy. The median age at PVR was 29 years (range, 12 to 66 years). The mean time between initial surgery and PVR was 23.4 years. No death occurred. We observed an improvement in New York Heart Association functionality, rhythm regression disturbances, a reduction in right ventricular volumes and an increase in left ventricular function. For the moment, without any evidence for survival improvement, we keep this operation for symptomatic patients only.

Bioprostheses are also used in our centre for the repair of pulmonary agenesis in older children

In discussions of future perspectives, the most frequently described innovative non-invasive approach is percutaneous PVR with the Melody Transcatheter Pulmonary Valve (Medtronic Inc., Minneapolis, MN, USA). This technique is actually limited to a calcified pulmonary prosthetic conduit not exceeding 22 mm in diameter [47], however, and is not for patients with repaired tetralogy of Fallot with a clearly dilated or even aneurysmal pulmonary artery requiring a device of larger diameter. Bioprosthesis at first PVR supplies a later percutaneous PVR due to the stented bioprosthesis. Tissue-engineered pulmonary valves may be the answer, but that answer seems far away.

Without prostheses

In fact, to avoid several reinterventions, some authors recommend—as in truncus arteriosus—RVOT reconstruction with a partially competent monocuspid valve [48,49] or even without any valve (54), but long-term data are still lacking. The late incidence of reintervention for RVOT obstruction or pulmonary valve implantation remains to be determined [50].

Tricuspid valve replacement

Of all the cardiac valves, the tricuspid is the least common to require replacement, making up less than 2% of VRs in the adult population. Tricuspid valve replacement (TVR) is even less frequently required in children. Irreparable Ebstein’s tricuspid valves (TVs) and TV endocarditis are the two most frequent indications for TVR in children. In the largest single-centre report of TVR, the Mayo Clinic (Rochester, MN, USA) performed more than 323 TVRs [51]. The need to replace the TV in their large Ebstein’s experience was greater than 50%. TVR was performed when TV repair was not feasible. The surprising conclusion of this sentinel report was that older children requiring TVR with Ebstein’s anomaly fared better than children requiring TVR for other disease entities. Bioprosthetic valves fared better in the TV position than the same bioprosthesis in other cardiac positions. Kiziltan et al. [51] reported on 158 consecutive patients requiring TVR for Ebstein’s anomaly at the Mayo Clinic over a 25-year period. Follow-up of 149 patients who survived 30 days ranged up to 17.8 years, with a mean of 4.5 years. Survival was 92.5% at both 10 and 15 years, with
nine late deaths. Freedom from bioprosthesis replacement was 97.5% at 5 years and 80.6% at both 10 and 15 years. Significant differences were noted with regard to freedom from reoperation for bioprosthesis in the tricuspid position compared with all other cardiac positions (Fig. 8) [8]. In addition, freedom from reoperation was lower for bioprostheses than for mechanical prostheses in the tricuspid position. Further support for bioprostheses in the tricuspid position has been provided by Guerra et al. [52], who reported a 14-year follow-up on 45 patients at a single institution. Thirty-eight of these 45 patients also had other valves replaced in other cardiac positions simultaneously with their TVR. Morphological examination of explanted porcine bioprostheses showed that those implanted in the tricuspid position had lower degrees of calcification and less severe structural changes than those simultaneously explanted from the mitral position. Overall, actuarial freedom from structural deterioration at 14 years for the bioprosthesis was 68% in the tricuspid position.

Shrestha et al. [53], in a study of homograft TVR, found that partial replacement of the TV using a homograft provided favourable in-hospital and mid- to long-term clinical outcomes for patients with a variety of causes and age groups, despite a slightly complex surgical technique compared with prosthetic VR. This procedure might be useful, particularly in treating active bacterial endocarditis or young patients, in centres at which homograft tissue is available.

Finally, Husain and Brown [8] recommend that the valve placement should be cephalad to the coronary sinus, the atroventricular node and occasionally the right coronary artery, to decrease the risk that the prosthetic sewing ring or struts will compromise these structures; great care should be taken to ensure that the struts of the bioprosthesis straddle the area of the membranous septum and conduction tissue; the valve should be seated into position with the heart beating to observe rhythm disturbances; and concomitant procedures, such as a right-sided Maze procedure, can and should be performed at the time of TVR if indicated.

Conclusions

VR in children is a complex issue because of initial valve and patient size, anatomy and growth potential. A multitude of opinions and surgical experiences exist. Today, the valve choices that seem without controversy are bioprosthetic VR of the TV and Ross AVR for the aortic valve. On the other hand, PVR choice remains controversial. Several good options are available for most valvular positions when VR is required. MVR in small children remains the most problematic. The Ross procedure produces excellent results in the aortic position and has shown promise in the mitral position in selected patients. The PA offers a potential durable option in both of these positions. Mechanical VR options do exist for children; however, chronic anticoagulation makes MVs less desirable. Future advances with tissue-engineered heart valves for all positions and new anticoagulants may change the landscape for VR in the paediatric population.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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References

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