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Review Article

Children with Congenital Disease Amenable to Aortic Valve Repairs: Lessons for Management and where do these Techniques Currently Stand?

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Abstract

Aortic Valve Repair (AVRep) appeared to be the first considered in the armamentarium to treat AV disease in children of various ages for the last two decades because of its lifelong durability and reproducibility. Several advantages of AV Rep over techniques of Aortic Valve Replacement (AVR) are identified. Surgical repair strategy should be individualized to the age of the patient depending on aortic root growth potential. New repair techniques developed by innovative surgeons are gradually becoming adopted.

Keywords: Aortic valve; Aortic valve neocuspidization; Aortic valve repair; Children; Reintervention

Introduction

The Aortic Valve (AV) can be commonly affected in children by several disease pathologies, frequently requiring multiple palliative intervention. Surgical repair for congenital Aortic Stenosis (AS) with cardiopulmonary bypass was first described by Lillehei, et al. and colleagues in 1956 and was the only treatment option for three decades [1]. Surgery initially had a high mortality rate, over 50% in the 1980 to 1990s [2]. During the 1980s and 1990s, the development of AVRep in children was veiled by the pre-eminence of competing Balloon Valvuloplasty (BV) and popularized Ross procedure. In 1983, BV was developed and quickly became the treatment of choice in many centers, seen to be a safer, less invasive alternative to surgery [3]. Disappointingly, because of the progressive dilatation of pulmonary autografts, a quarter of these patients may require reintervention on the autograft within 2 decades of the Ross procedure. Fortunately, Since the late 1990s, AVRep techniques have been refined and have become feasible in adult patients with a 10-year freedom from reoperation of more than 80% [4,5]. As with mitral valve surgery, these techniques with their lessons learned are being increasingly adopted daily in children with promising mid-term results and more recently long-term results (Table 1). However, there are two basic weaknesses in extending the adult experience in AVRep in children. First, extensive external annular reinforcement in small children would prevent growth. Second and most importantly, the age at presentation is a marker of severity and the pathology in adults is far less severe than in children. Nevertheless, any Aortic Valve (AV) operation in children (repair, Ross or mechanical replacement) is a palliation and reinterventions are frequent. One justification for use of AV repair is the ability to delay AV replacement techniques to an older age. The ultimate goal of these AVRep or reconstruction techniques is to provide the best survival, minimal reintervention risk, and high quality of life. From this point of view, the purpose of this article is to analyze the current surgical strategies and techniques of treating AV disease and unsolved repair related issues in children.

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What is the primary procedure of choice in children with aortic valve stenosis?

Treatment of congenital AV stenosis usually requires multiple interventions. The ultimate goal of treatment should result in the adequate relief of obstruction while minimizing significant regurgitation. Both BV and Surgical Aortic Valvulotomy (SAV) have been widely applied, but the primary intervention for congenital AV stenosis in children still remains controversial [6].

In 2015, Prijic, et al. and colleagues reported a study that evaluated the long-term results of BV (n=39, 1.3 months to 17 years old) and SAV (n=23, 1.2 months to 15 years old) from 1987 to 2013. The freedom from reintervention rates were 71% for SAVs and 61% for BV in 10 years, but 42%for SAV and 23% for BV in 20 years [7]. These results reveal that the long-term outcome of SAV is better that that of BV, though the early results might look like less different. Not surprisingly, freedom from Aortic Valve Replacement (AVR) for BV (32%) is lower than that for SAV (53%). Butno matter what type of surgery we choose, congenital AS carries the lifelong risk of reintervention.

In 2015, Soulatges and colleagues analyzed 93patients (1 day to 18 years, 2,4 years on average) who underwent BV as the primary treatment from 1992 to 2012 [8]. The long-term survival rate was 88,2% in whole cohort. The freedom from reintervention rate was 58% (54% in neonates and 65,6% in infants) with the mean follow-up duration of 11,4 years and the freedom from surgery was 66% (58,5 in neonates and 75,8 in infants). The applied as the primary treatment

in infants and children. While in 2006, Miyamoto and colleagues reported a freedom from reintervention of 85,1%, 78,0%, and 53,5% at 5, 10, and 15 years respectively in patients who underwent primary SAV due to the critical AS aged less than 3 months between 1983 and 2003 and they concluded that SAV was the most appropriate strategy to treat neonates and young infants with critical AS.

Authors	Journal	Year	Number	Age	Technique	Material	Mortality	Follow-up	Results	Freedom from re-intervention
Hasaniya et al (Loma Linda)	JTCVS	2004	21	Mean 8.1 years	Nocuspextension	-	None	Mean 5.3 years	11% residual moderate AR postoperatively	47% reoperation at mean 28,3 months postoperatively
Tweddell et al (Milwaukee) [29]	JTCVS	2005	147	Mean 6 years	Simple repair (valvotomy, commissurotomy + thinning)	-	Early: 15 (10.2%), 2 latedeaths	Mean 2.9 years	Moderate AR:27%, severe AI: 6%, mean AS gradient: 20 mm Hg	96% at 5 years, 86% at 10 years
Odimetal (LosAngeles)	ATS	2005	31	Mean 25.5 years	Multiple ıoleaflet extension in all	GTAP	Early: one, no late deaths	Mean 25 months	Moderate AR in 6%, moderate AS in one patient	Patients< 18 years:100% at 2 years
Alsoufi (Toronto)	ATS	2006	22	Mean 11.4 years	Multiple ıoleaflet extension in all	GTAP	No early or late deaths	Median 1.7 years	Moderate AR in seven (32%) patients, median postoperative gradient 29 mm Hg, 7 (32%) > 30 mm Hg postoperatively	80% at 2.5 years
Hawkins (Salt-LakeCity)	JTCVS	2006	54	Mean 8.5 years	Multipleincluding leafletextension	GTAP	Early: one (1.9%), no late deaths	Mean 76 months	Reoperation: 13/54 (23%) at 48 months postrepair, two patients with moderate severe AR	68% at 5 years, 58% at 10 years
McMullan (Melbourne)	JTCVS	2007	21	Mean 12.6 years	Multiple ıoleaflet extension in all	GTAP	No early or late deaths	Mean 34.7 months	Early reoperation: two, late reoperation: two, moderate AR: 4 of 21 (25%), severe AS: one	Reoperation > moderate AI: 75% at 36 months
Quader (Cleveland)	Heart, Lung & Circulatio-mon	2006	56	Mean 13.4 years	Multiple, cusp extensionin 14%	GTAP	Early death: one, Late death: two	Median 37 months	Aortic stenosis group, n j7: 4 (57%) _ moderate AS, AR group, n j24: 3 (12.5%) _ moderate AR, 6(11%) requiredreoperation	90% at 1 year, 80% at 5 years
Bacha (Boston) [23]	JTCVS	2008	81	Median 8.6 years	Multipleincluding leafletextension	GTAP	Three (3.7%) early and late combined	Median 4.5 years	Rerepair: three, valve replacement: 25 (31%), balloon aortic valvuloplasty: 8 (10%), in the remaining patients-moderate AR: 21 (38%) and moderate AS: 14%	91% at 1 year, 63% at 5 years
D'Udekem (Melbourne) [21]	JTCVS	2013	142	Median 9 years	Leafletextension, n = 96	GTAP	Twoearly (1.4%), onelate	Mean 3.4 years	Moderate AR: 23 (19%), severe AR: 1, moderate AS: 12 (10%), coronary ischemia, n j5 (early death 2, ECMO 1, on-table revision 2	80% at7 years
Kandakure, et al. (Liverpool)	WJPCHS	2014	39	Median 5.5 years	Multiple including leaflet extension, neocommissure creation in all	GTAP	Early: 3 (7.7%),	Median 12.7 months	Moderate AR: 2 (5.1%), Moderate AS: 3 (7.7%)	95% at 3 years
Vergnat et al (Sankt Augustin)[32]	EACTS	2017	193	Median 9 years	Multiple including leaflet extension, replacement and neocommissure creation in all	GTAP	Early: 1 (0.5%), 2 latedeaths(1%)	Mean 5.1 years	Moderate AR: 13 (27%), Moderate AS: 7 (5,7%)	89%, 70%, 57% at 1, 5 and 7 years postoperatively
Fraser et al (Baltimore) [41]	JTCVS	2019	100	Median 13.6 years	VSRR	-	2(2%) Peri-operative, 7 latedeaths(7%)	Median 4.7 years	Moderate and greater AR: 9 (9%)	100%, 88.5%, 70.4% at 1, 5 and 10 years postoperatively

Table 1: Aortic valve repair in the pediatric population for congenital aortic valve disease[a].

Abbreviations: ATS: Annals of Thoracic Surgery; JTCVS: Journal of Thoracic and Cardiovascular Surgery; GTAP: Glutaraldehyde-Treated Autologous Pericardium; AR: Aortic Regurgitation; AS: Aortic Stenosis.

[a]Recently published series with mean or median follow-up less than ten years.

Collectively, these data suggest that the outcomes of BV in younger patients were no better, if not worse, than those of SAV [9]. In reference to a recent meta-analysis [10], we can intuitively see that SAV and BV had similar early mortality, but the long-term outcomes (the freedom from reintervention) of SAV were superior overall and in infant alone (P<0,001).

The Ross procedure: the ultimate operation?

AVR with a pulmonary autograft, the Ross procedure, is widely used in children. Given the advantageous features of the autograft conduit for children (absence of anticoagulation, no age limitation, growth potential), Ross procedure became the operation of choice in children. The German Dutch Ross registry shows a 10-year freedom of reoperation in excess of 90%, far superior to any report on AoV repair for Aortic Regurgitation (AR) in children that is consistent with a report comprised a 10-year freedom from reoperation of 95,2% in a series of 31 neonates and infants [11,12]. Nevertheless, there is a growing body of evidence in the paediatric field that dilatation and failure of neoaorta occur in up to 28% at median follow-up duration of 5,2 years after Ross procedure [13].

In the study from Children's Hospital of Philadelphia, 97% of the children had a neoaortic root z-score of >4 at six years and 60% had moderate or greater neoaortic regurgitation [14].

Techniques preventing this drawback (modified Ross) have developed but can only be applied in a mature child. Performing the AVRep would allow for aortic root growth and the postponement of the Ross procedure to a time when such modified procedure or if possible [15], secondary AVRep can be used. In the Vergnat, et al. report, of the 48 patients who eventually required AVR, the median age at repair was 8,0 years, and replacement could be postponed at a median age of 12,0, which allowed in some patients the use of supportive Ross techniques [16]. As described later, older age may also allow consideration of further techniques for recurrent repair (such as leaflet replacement) that could have not been used in younger patients because of limited durability.

Aortic valve repair techniques: current development and results

With the development of valve repair techniques, AVRep shows several advantages over replacement. On the one hand, repair can be performed in almost all ages and utilizes native tissue, do not require anticoagulation, and has a low risk of endocarditis, calcification or thromboembolism. Furthermore, valve repair stabilizes Left Ventricular (LV) dimensions, hemodynamics and results in symptomatic improvement, and various strategies can be utilized as the patient grows. Surgical repair of AV can be classified as simple or complex. Simple methods include valvulotomy, valve debridement, commissurotomy, valve suspension, and commissure suspension. Complex AVRep methods include cusp extension, leaflet replacement, neocommissure creation. Extensive valve debridement, nodular fibrosis resection, fused commissures opening and thickened cusp areas thinning are usually involved in the treatment of stenotic AV disease [17].

Recently, there have been a number of publications describing advance repair techniques with favourable results for congenital AV pathology, especially bicuspid AV in adults (pioneered by El Khoury,et al. [18] and Schaffers [19]). The latter evolvement and demonstration of the reliability and superiority of repair in the mitral domain and emergence of Ross paediatric patients in their second postoperative decade, showing up to 40% autograft failure at 15 years [20], superior results of SAV over BV in diverse children age groups. All these events renewed the interest in pursuing valve repair in children.

For these reasons, since 2000, few centres have developed a repair-oriented policy for AV disease in children. Among them, the Melbourne centre has developed world-renowned expertise [21]. However, long-term data are scarce because of the newness of the technique and the paucity of paediatric cohorts.

Surgical repair of AV can be classified as simple or complex. Simple methods include valvulotomy, valve debridement, commissurotomy, valve suspension, and commissure suspension. Complex AVRep methods include cusp extension, leaflet replacement, and other valve reconstruction. Ideally, AVRep would relieve the hemodynamic burden on the LV, permit growth, provide the tolerance against infectious endocarditis, avoid AVR and the need for anticoagulation. Thus, the implementation of AVRep is extremely broadly favored that allow the largest proportion of patients to have the smallest number of reinterventions over their lifetime. Regarding the Poncelet, et al. data, the latter event contributed to the expansion of the use of AVRep air. The enrollment pattern for AVRep, Ross procedures and AVR over four decades is illustrated in Figure 1. So, Poncelet, et al. reported the freedom rates from AVR at 5, 10 and 15 years of 90, 81,1 and 74,8%, respectively. compare favourably to other contemporary studies [22]. Indeed, Bacha, et al. [23] reported 5- and 10-year rates of freedom from AVR of 72% and less than 50%, respectively.

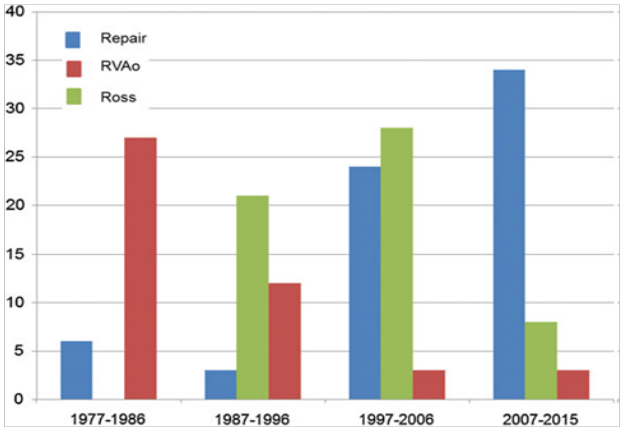

Figure 1: Number of aortic valve (AV) repair (blue bar), AV replacement (red bar) and Ross procedures (green bar) by decade. Brussel experience.

Khan, et al. reported a 5-year rate of freedom from AV reintervention of 75% (excluding the truncal repair group) [24]; and d'Udekem, et al. [21] recently reported 90 and 50% freedom from AVR at 5 and 10 years for their entire cohort of repair, which included about 100 paediatric patients older than 1-year-old. The frequency of AVRep is also, as in Brussel experience, favored in contrast to BV and Ross operation (Figure 2). The stability of the repair in our study was satisfactory with 90, 72 and 72% freedom from AV reintervention at 5-, 10- and 15 years, respectively. One of the reasons for this improved longevity might reside in the fact that in Melbourne experience, it was rarely used leaflet extension as a tool to obtain a competent AV.

Only 10 patients (15%) underwent leaflet extension in our 38 years of experience. In comparison, leaflet extension was used in 30% of the patients in the Melbourne experience and in 80% of the Boston experience [21,23]. Indeed, the subgroup of patients in Melbourne without the leaflet extension technique had a much higher freedom from AV reintervention. Also supporting this hypothesis, Wilder, et al. [25] demonstrated a nearly 50% valve-related reoperation rate in their series of older children when they used leaflet extension, with a mean time to re-repair/replacement of 4,4 years. Recently, Vergnat, at al. showed the favorable long-term results as a freedom rate from reoperation at 1, 5 and 7 years of 89% (95% CI, 85-94%), 70% (95% CI, 63-78%) and 57% (95% CI,47-66%), respectively.

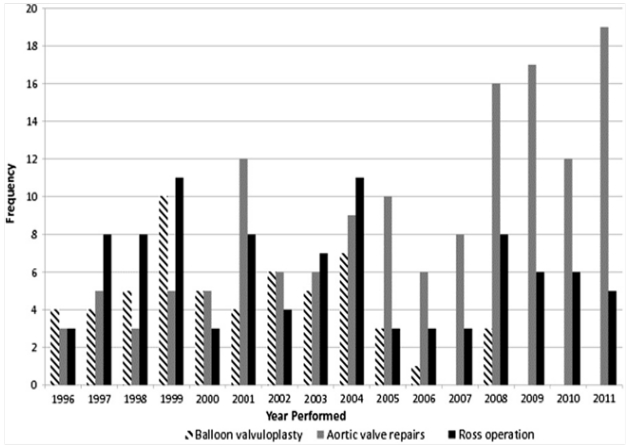

Figure 2: Aortic valve procedures. Melbourne experience.

The influence of the left ventricle preservation on valve repair results

Although efforts should be oriented to maintain the native valve in younger children as long as possible, preservation of LV function is also critical to this strategy. Indeed, LV failure is consistently associated with death in the Ross procedure [26]. For some authors, AVRep may prolong LV exposure to pressure or volume overload and thus jeopardize future outcome for these patients [27]. Thus, preserving the native valve as long as possible, without affecting LV function, leaves all other options still valid without limitations (supported Ross/mechanical valve and even recurrent repair).

Are aortic stenosis and aortic regurgitation equal with their post-repair prognosis?

Surgery for congenital AS in children generally involves commissurotomy plus thinning of the adherent fibrous tissue on the leaflets. These repairs are durable and show reoperation free survival in excess of 85% at 10 years [28,29]. Durable relief of regurgitation on the other hand, is a far greater challenge and here the results in children are uniformly poor with less than 50% freedom of reoperation at 8 years in several single institution series [29-31]. A number of techniques have been applied including leaflet suspension, restructuring and augmentation of the commissures with pericardium and leaflet extensions. Despite the ability to yield excellent immediate results the repairs lack durability and within too short a time, regurgitation happens, as repairs for regurgitation require extensive use of additional materials.

Aortic root growth and aortic valve geometry-based strategy

Patient growth has a major impact on repair durability as it varies in various children ages. The growth potential of the AV complex can be considered in 2 directions: vertical (from cusp nadir to Arantius nodula) and horizontal (inter commissural). Regarding to growth of the AV and patients' ages, the whole repair procedures are subdivided in 2 variations:

- Unrestrictive procedures (commissurotomy shaving or neocommissure creation) maintain growth potentials in both directions
- Restrictive procedures (leaflet replacement and extension) restrain intercommissural growth Basing on Vergnat et al. data, a restrictive procedure in small children (especially under the age of 1 year) leads to a reoperation sooner ratherthan later. In young patients (<10 years), neocommissure creation is promising; 2 other techniques (commissurotomy shaving or leaf let replacement) are equally effective. In older patients (>10 years), all techniques except leaflet extension offered 80% freedom from reoperation at 8 years of follow-up [32]

Stenosis as the primary indication was present in about 2/3 of the patients. The results show a 80% freedom of reoperation at 8 years of follow-up, entirely consistent with the previous studies cited above. The end point in the study by Vergnat, et al. is freedom from reoperation. While this end point is widely accepted, reoperation is a measure of physician behaviour and is not always a reflection of the physiologic burden on the LV, that is, recurrent stenosis or regurgitation impacting on the success of repair.

Valve morphology may influence on outcomes with tricuspid morphology providing a protective effect against reoperation and, more importantly, against replacement [32]. This effect may be due either to the underlying morphology, to a better geometrical configuration favouring durability or to the maintenance of the further possibility of re-repair (leaflet rereplacement instead of valve replacement).

Aortic valve repair by using patch material

As noted above, regurgitation repairs fail in not too distant time, since repairs for this type of disease demand extensive use of additional materials either xenograft patches or fixed autologous pericardium. In addition, suture lines are required to flex repeatedly and repair forces are commonly concentrated into a small region, resulting in structural failure. These materials are subject to rapid degeneration due to fibrosis with retraction and calcification. In primary repairs with a usage of treated autologous pericardium reoperation shows 2 main mechanisms of failure: restrictive motion leading to late reoperation and excessive motion leading to early reoperation. Though experience and reports in the literature have accumulated, the debate on the optimal material is still ongoing, especially given the disparity of data (children and adults mixed), the small number of pediatric patients and the new materials.

Outcomes of the aortic valve neocuspidization-Ozaki technique in children

AV reconstruction was first reported by Dr. Duran, et al. His procedure attempted to reproduce tailored valves as much as possible [33]. They used three consecutive bulges of different sizes as templates to

guide the shaping of the pericardium, which was made according to the dimensions of the aortic annulus. The pericardial leaflets were sutured to the AV remnant or to the annulus. Duran shared the experience of 51 patients who underwent Duran procedure with the survival rate of 84,53% at the follow-up of 60 months, and 72,59% of the patients was free from any event after the operation. He concluded that early outcomes encouraged us to reconstruct the AV by using autologous pericardium.

Aortic Valve Neocuspidization (AVNeo) was developed by Ozaki nearly a decade ago [34]. The Ozaki procedure consists of using glutaraldehyde-treated autologous pericardium to replace aortic valve leaflets but improves significantly in measuring the diseased valve and using templates to cut fixed pericardium compared with the Duran procedure. The suture line forces are distributed over a much a larger area, and the new leaflets have a large area of coaptation, further relieving stress on the repair. As no foreign material is required, postoperative anticoagulation is unnecessary, and the AVNeo was thus considered as valve repair rather than replacement [32]. AVNeo has become more widely adopted with the advent of leaflet templates, thereby making the technique more standardized and reproducible. AVNeo achieved excellent result in large series of patients, but pediatric cases accounted a small proportion and further follow-up is needed particularly in the pediatric population. In this setting, The Boston Children's Hospital experience with AVNeo began in 2014. Regarding a report of their experience, 57 patients with a median age of 12,4 years underwent Ozaki procedure [35]. Twenty-four patients had AR, 6 had AS, and 27 patients had mixed disease. Four patients had truncus arteriosus. Thirty-four patients had prior AV repairs and 5 had replacements. Autologous, Photofix®, and CardioCel® bovine pericardium were used in 20, 35, and 2 patients. Eight patients underwent aortic root enlargements and 20 had non-coronary sinus enlargements. There were no hospital mortalities or early conversions to valve replacement. There were no differences in outcomes based on operative indication. At median follow-up of 8,1 months, 96% and 91% of patients had less than moderate regurgitation and stenosis, respectively [35].

In 2015, Mazzitelli, et al. reported their experience with 3 pediatric AVNeo procedures in patients 5, 11, and 15 years of age. Tissue-engineered bovine pericardium (Cardiocel®) was used to create cusps and there were no reoperations at early follow-up [36].

In addition, a more uniform repair technique amenable to a variety of anatomic situations may simplify our approach and achieve consistent results. One persistent weakness is the use of devitalized pericardium that will predictably degenerate in children. Perhaps non-bioprosthetic material may be considered such as 0,1 mm polytetrafluoroethylene. While it will not grow, it is not subject to degeneration or neointimal in growth and could prove to be more durable.

Congenital aortic valve diseases associated aneurysm of the proximal aorta and aortic sinuses: The supplement for guideline treatment

Aortic root aneurysms are rare in children that are usually associated with connective tissue diseases or conotruncal congenital heart anomalies**.** The traditional surgical approach for these pathologies has been to replace the aortic root with concomitant AVR using a valve-and-graft composite, the so-called Bentall procedure. Despite the yielded low mortality and low morbidity at follow-up, AVR with mechanical valve mandates lifelong anticoagulation or potential need for reoperation because of degeneration of placed bioprosthetic valve. Thus, Valve-Sparing Root Replacement (VSRR) has evolved over the past few decades to improve the quality of life for patients requiring these procedures through the pioneering work surgeons such as David and colleagues [37], David and Feindel, et al. [38], and Sarsam and Yacoub, et al. [39]. VSRRis an appealing surgical approach in children with aortic root aneurysms because it allows for avoidance of the problems associated with long-term anticoagulation, carries a low risk of thromboembolism or endocarditis, and has shown promising intermediate-term outcomes. Given the absence of predictors of dissection or rupture in paediatric patients presenting with aortic root aneurysms [40], absolute indications for intervention in children remain elusive. Nevertheless, VSRR is undertaken to mollify further dilatation and hamper potentially fatal complication as rupture. Recently, Fraser et alreported an experience that highlighted a freedom from reoperation of 100%, 88,5% and 70,4% at 1, 5 and 10 years, respectively, after the 100 consecutive valve-sparing root replacement in children with a median age of 13,6 years [41]. Their data also attested to the durability of the reimplantation technique that is superior when compared with remodeling technique by stabilizing the aortic root, improving hemostasis. Moreover, of the 84 patients undergoing a reimplantation procedure, 4 (4,8%) underwent late AVR versus 5 (31,3%) of the 16 patients who underwent a remodeling procedure (p= 0,001) [41].

Institutional experience and patient selection are known as crucial factors and relate to outcomes after VSRR, particularly in children. Accordingly, the Johns Hopkins Hospital elaborated the well-known guidelines for aortic root replacement that are driven on the basis of the presence of connective tissue disorders and the disease subtype or severity of disease taking into account of multiple risk factors [41] (Table 2). It should be noted that there have been instituted several contraindications for VSRR in children with connective tissue disorders as significant leaflet fenestration and asymmetry, acute aortic dissection in unstable patients, bicuspid aortic valve (BAV) with extensive calcification, severe prolapse, marked fenestrations.

Diagnosis	Indications for VSRR in Children
Marfan Syndrome	Maximum diameter>5,0 cm or increase of>0,5 cm/y Diameter of 4,5-5,0 cm if: Family history of or rupture Aortic valve regurgitation Need for mitral valve repair and aortic root 4,0-5,0 cm
LDS LDS Types I and II	Maximal diameter of>3,5-4,0 cm or Z score>3 Increase in diameter of>0,5 cm/y Severe craniofacial features
LDS Type III	Maximal diameter of>4,0-4,5 cm or Z score>4 Increase in diameter of>0,5 cm/y
LDS Type IV	Maximal diameter of>4,5 cm or Z score>4 Increase in diameter of>0,5 cm/y
Bicuspid Aortic Valve	Maximal diameter>5,5 cm
Non Syndromic Thoracic Aortic Aneurysms	Maximaldiameter>5,5 cm

Table 2: Current guidelines for aortic root replacement at the Johns Hopkins Hospital.

Abbreviations: VSSR: Valve-sparing root replacement; LDS: Loeys-Dietz syndrome.

Bicuspid aortic valve associated aortopathy: A complication with high incidence need to pay more consideration

BAV is the most common congenital cardiac malformation and is present in 1% to 2% of thegeneral population [42]. Dilation of the aorta is a frequent complication in patients with BAV and the aorta in this population grows more rapidly compared to the normal population [43]. Although the majority of young patients with BAV have mild dilation, the rare patient can have progressive dilation and is prone to risk for morbidity and premature death [44].

Ruzmetov, et al. demonstrated that the R/N fusion of BAV was strongly associated with ascending aorta dilatation [45], whereas patients with R/L subtype were more likely to have aortic root dilataion as measured by change in Z-score over time. In addition, patients with R/N fusion presented at a younger age and were more likely to have AS. The latter data is consistent with a report of Fernandes and his colleagues [46]. Revealing the association aortic dilation with BAV, Schaefer and associates proposed an integrated classification system based on both leaflet fusion and aortic root shape [47]. The authors found that patients with R/L leaflet fusion most commonly presented with type "N" (normal shape), whereas those with R/N leaflet fusion were more likely to have a type "A" (ascending dilation) or type "E" (effaced root) root anatomy. The latter finding also was found in in a series of Children's Hospital of Illinois [45].

Some studies found that moderate or greater AR was associated with higher aortic root and ascending aortic diameter Z-scores in children with BAV [48,49]. Nonetheless, it is unclear whether dilation entails regurgitation, regurgitation begets dilation, or both. Some authors have considered haemodynamic causes that could account for the AR contributing toaortic dilation such as increased wall stress due to increased stroke volume from AR [48]. Other authors have suggested that aortic dilation likely precedes AR with the progression of aortic dilation causing poor coaptation of the aortic valve leaflets that leads to AR [50].

Despite considerable controversy, some believe that the vascular complications of BAV are not secondary to valvular dysfunction and can manifest in young adults without significant AS or regurgitation [51,52]. Nistri and colleagues determined that more than 50% of young patients with normally functioning BAVs have echocardiographic evidence of aortic dilation [51], whereas in a series of Children's Hospital of Illinois [45], 33% of young patients with BAV have evidence of dilation (z score >3).

Conclusion

Commissurotomy with thinning appeared to be approved as regards AS underlining its durability and real-world applicability. Allowing the patient to grow to an age when more definitive solutions are available, surgical AVRep strategy should be individualized to the age of the patient. AVRep can achieve intermediate and long-term results without affecting LV function that may be a reasonable primary choice in comparison with AVR in children, especially if the alternative is prosthetic valve replacement that requires extensive annular enlargement. Ultimately, AVRep should reduce the use of replacement procedures in the growing child. Future work in this area should be directed to developing more consistent techniques and better materials that will result in more durable repairs. In children, for whom the surgical options including AVRep numerous techniques may be limited, AVNeo can be considered as an alternative method for application. Some studies suggest that BAV morphology may be associated with patterns of aortic dilatation and valve dysfunction. Recognition of these differences may eventually be helpful for echocardiographic surveillance and potential medical intervention.

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