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Ascending aorta and aortic root replacement (with or without valve sparing) in early childhood: surgical strategies and long-term outcomes

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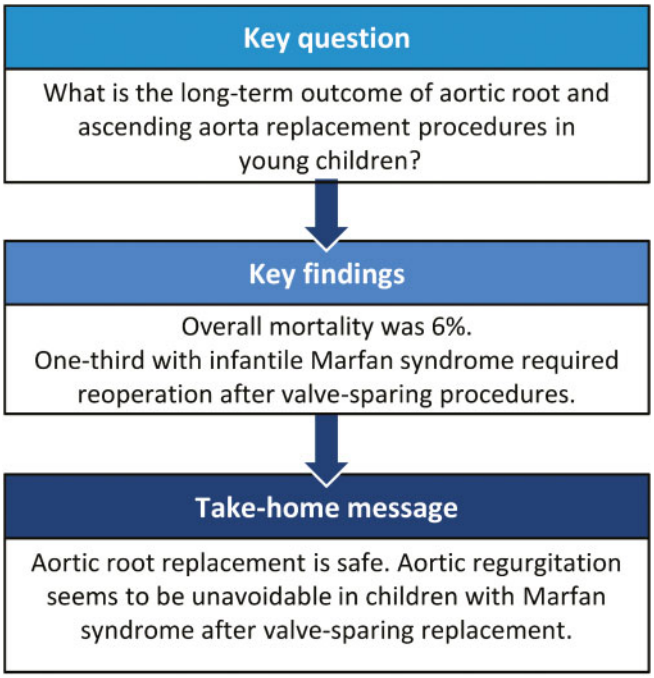

Abstract

OBJECTIVES: Aortic root and ascending aorta replacements (AARs) are rarely required in the paediatric population. We report here a series of AAR performed in young children using different surgical techniques.

METHODS: Between 1995 and 2017, 32 children under the age of 10 years (median age 5.4 years) underwent AAR procedures at our institution. Twenty-two (69%) had a connective tissue disease (infantile Marfan syndrome or Loeys–Dietz syndrome). We performed 11 AAR using a composite graft with a mechanical prosthesis and 21 valve-sparing procedures (10 Yacoub operations and 11 David operations). Median follow-up for operative survivors was 7.7 years (interquartile range 4.2–12.8 years).

RESULTS: The cardiac-related early mortality rate was 6%. Patient survival was 91% at both 1 and 10 years. Eleven survivors (38%), all with a status of post-valve-sparing procedure, required an aortic root reintervention with an aortic valve replacement after a median interval of 4.2 years. Interestingly, only patients with infantile Marfan syndrome tended to be associated with risk of reoperation.

CONCLUSIONS: Aortic root and AARs are safe in young children whatever the surgical procedure. Aortic valve-sparing procedures show good long-term results except in children with infantile Marfan syndrome whose ineluctable aortic annulus dilatation or aortic valve regurgitation requires reintervention after a short period.

Keywords: Aortic root replacement • Ascending aorta replacement • Aortic root dilatation • Children • Bentall • Valve-sparing procedures • Infantile Marfan syndrome

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INTRODUCTION

Aortic root and ascending aorta replacements (AARs) are rarely required in the paediatric population and are generally performed in patients with connective tissue defects, such as Marfan syndrome (MS) and Loeys–Dietz syndrome (LDS). These conditions in turn cause progressive and serious aortic root aneurysms with a high risk of dissection or rupture. AAR may also be of benefit to children who have undergone neonatal repair of complex congenital heart diseases (CHDs) involving the outflow tract such as the Ross procedure and the arterial switch operation.

Various operative techniques for AAR have been developed since the first description by Bentall in 1968 of total AAR using a composite mechanical valved graft [1]. Among these techniques, the remodelling technique (Yacoub procedure) [2] and the reimplantation technique (David procedure) [3] are the preferred options in young children since they allow to preserve the aortic valve. These recent valve-sparing procedures are very attractive because they do not necessitate life-long anticoagulation treatment and have demonstrated good results in adults [4–6]. However, they still need long-term outcome assessment when performed in young children and need to be compared with outcomes for the Bentall operation (composite mechanical valve graft replacement).

This study reports results from the long-term follow-up of young children who required an AAR procedure at our institution. We decided to focus this review on patients younger than 10 years of age because (i) surgical management and results in post-pubescent children are similar to those observed in the adult population; and (ii) it has been shown that children with MS diagnosed before 10 years of age represent a particularly high-risk group [7].

PATIENTS AND METHODS

Patient population

Between 1995 and 2017, 32 children under the age of 10 years (21 boys and 11 girls) underwent an AAR at our institution. We reviewed retrospectively their demographic, clinical, echocardiographic and surgical records. We considered the surgical indication and findings from the last preoperative echocardiographic assessment. Adverse events were also noted, including postoperative complications (acute bleeding, atrioventricular block, low cardiac output, pericardial effusion), need for reoperation and death. Data were collected either during a medical visit or *via* a telephone interview with patients or their referring cardiologist.

The median age of the cohort was 5.4 years [interquartile range (IQR) 3.3–7.4 years] and the median weight was 18 kg (IQR 12.2–23.9 kg). Twenty-two of the patients (69%) had a connective tissue disease [infantile Marfan syndrome (IMS) or LDS] and 7 (22%) had undergone an operation as a neonate for CHD [transposition of great arteries ($n = 2$), critical aortic stenosis ($n = 2$), common arterial trunk ($n = 2$) and double outlet right ventricle with interrupted aortic arch ($n = 1$)]. The 3 remaining patients had an isolated bicuspid aortic valve. Seven patients with IMS or LDS also had a bicuspid aortic valve.

All the patients with IMS ($n = 15$) were diagnosed in infancy with a clinical phenotype. In 10 of these patients (67%), we confirmed the diagnosis by *FBN1* gene sequencing (9 had a typical mutation between exons 24 and 32; 1 had an atypical deletion in exon 43). All the patients with LDS ($n = 7$) had documented *TGFBR1* or *TGFBR2* mutations.

Surgical procedures

The decision to perform AAR was made by considering the severity of the aortic root dilatation and/or the rate of increase in root diameter.

Prior to AAR, 10 patients (31%) had undergone at least 1 cardiovascular surgery: mitral repair in 2; aortic valvar procedure in 6 (commissurotomy, pulmonary autograft transfer, valve repair, mechanical prosthesis placement); arterial switch operation in 3; aortic arch repair in 2; patent arterial duct closure in 1; and common arterial trunk repair in 2.

We performed AAR through a median sternotomy using standard cardiopulmonary bypass with aortic and bicaval venous cannulation. The valve-sparing remodelling procedure was performed, as first described by Sarsam and Yacoub [2], using a tubular Dacron graft into which the coronary arteries were reimplanted. The valve-sparing reimplantation procedure was performed using a straight-tube Dacron graft (David I) in which the native aortic valve was resuspended without a Valsalva graft [3]. One patient with moderate aortic insufficiency underwent a Yacoub procedure associated with the placement of an external ring to support the ventriculoaortic junction. The decision to perform a David or a Yacoub procedure was made according to surgeon preference, keeping in mind that the Yacoub procedure, while allowing for growth of the aortic annulus in young patients, may result in undesirable, excessive late aortic annulus dilatation.

Statistical analyses

Echocardiographic parameters were expressed both in absolute values and as scores indexed to body surface area (*z*-score according to the Dubois formula) [8].

Categorical variables were expressed as frequencies and compared using the Fisher's exact test. Continuous variables were summarized as medians with IQR and were compared using the Kruskal–Wallis test. When the Shapiro test established the normal distribution of the variables, we used the analysis of variance test. We performed Cox proportional hazards regression analysis to determine risk factors for reoperation. Given the low numbers of both patients and outcome events, multivariable analysis was done but showed trends rather than significant values. Survival plots were made according to the Kaplan–Meier method, and comparisons between operative techniques for freedom from aortic root reoperation were performed using the log-rank test. A *P*-value of 0.05 was considered significant. All analyses were conducted using XL-Stats software (Addinsoft®, Paris, France).

RESULTS

Indications and operative techniques

The primary indication for an operation was aortic root dilatation in 30 patients (94%) and aortic regurgitation in 2. In these latter 2

Table 1: Patient characteristics and operative data according to aortic root replacement procedure

Characteristics	Bentall ($n = 11$)	David ($n = 11$)	Yacoub ($n = 10$)	*P*-value
Age (years)	7.6 (6.2–9)	5 (4.4–6.8)	3.6 (1.8–4.8)	0.045
Weight (kg)	22 (17.5–26.5)	18 (15.3–20.9)	11 (9.7–16.3)	0.10
Male, *n* (%)	9 (82)	6 (55)	6 (60)	0.42
IMS, *n* (%)	1 (9)	5 (45)	9 (90)	0.001
Preoperative AR >1+, *n* (%)	10 (91)	3 (27)	2 (20)	0.002
Preoperative aortic root (mm)	35 (32–36)	36 (35.6–43.8)	39 (33–40)	0.38
Preoperative aortic root *z*-score	+5.5 (+4.6 to +7.5)	+6.9 (+5.8 to +8)	+7.5 (+6.4 to +9)	0.12
Previous cardiac operation, *n* (%)	7 (64)	0	3 (30)	0.003
Cross-clamp time (min)	107.5 (84–140.8)	114 (101–120)	100 (83–119)	0.70
Aortic root graft (mm)	22 (22–23)	22 (20–23)	18 (15.5–20)	0.002

Quantitative values are expressed as medians (interquartile ranges).
AR: aortic regurgitation; IMS: infantile Marfan syndrome.

Table 2: Patient characteristics and operative data according to the underlying disease

Characteristics	IMS ($n = 15$)	LDS ($n = 7$)	Others ($n = 10$)	*P*-value
Age (years)	4.5 (3.1–5)	5.7 (1.9–6.8)	8.1 (6.5–9.3)	0.012
Weight (kg)	15 (10.5–20)	15.5 (10.8–17.5)	23 (18.5–26.8)	0.05
Male, *n* (%)	10 (67)	2 (29)	9 (90)	0.035
Preoperative AR > 1+, *n* (%)	3 (20)	3 (43)	9 (90)	0.002
Preoperative aortic root (mm)	39 (36–45)	35.5 (34–38)	34 (31.5–36.3)	0.16
Preoperative aortic root *z*-score	+7.4 (+6.4 to +8.4)	+7 (+6.3 to +8.4)	+4.9 (+4.8 to +6)	0.012
Previous cardiac surgery, *n* (%)	2 (13)	0	8 (80)	0.003
Aortic valve-sparing procedure, *n* (%)	14 (93)	5 (71)	2 (20)	<0.001
Cross-clamp time (min)	115 (96–138)	93 (87–109)	110 (85–145)	0.31
Aortic root graft (mm)	20 (18–22)	22 (20–22)	22 (22–23.5)	0.078

Quantitative values are expressed as medians (interquartile ranges).
AR: aortic regurgitation; IMS: infantile Marfan syndrome; LDS: Loeys–Dietz syndrome.

(1 after a transposition of great artery repair and 1 after a common arterial trunk correction), the decision to replace the ascending aorta was made to perform AAR to prevent redo surgery. Associated with the aortic root dilatation, 13 children had moderate or severe aortic insufficiency and 6 had significant (>mild) mitral regurgitation.

Prior to surgery, the median aortic root diameter at the sinus portion measured 36 mm (IQR 33–40.5 mm), with a corresponding median *z*-score of +7 (IQR +5.6 to +7.9). All the patients had a normal left ventricular ejection fraction.

Twenty-one patients (66%) underwent replacement of the aortic root with preservation of the native valve; the Yacoub procedure was performed in 10 patients (31.2%; age range 10 months–9.3 years); the David procedure was performed in 11 patients (34.4%; age range 11 months–9.7 years). Eleven patients (34.4%; age range 7 months–9.5 years) had an AAR using a composite graft with a mechanical prosthesis (median size 21 mm; range 17–23 mm). The most common graft size for AAR in this cohort was 22 mm.

Preoperative and operative data related to either surgical procedure or to the underlying congenital disease are shown in Tables 1 and 2, respectively. Overall, patients with connective tissue disorders were younger at surgery [4.6 (IQR 2.8–6.1) years vs 8.1 (IQR 6.5–9.3) years; $P = 0.003$] and exhibited greater aortic root dilatation [*z*-score +7.2 (IQR +6.2 to +8.6) vs +4.9 (IQR +4.4 to +6); $P = 0.003$]. Patients who had undergone the Yacoub procedure were operated on at younger ages than those who had undergone other procedures and consistently received smaller aortic root grafts. Composite graft implantation was the procedure of choice in children with moderate to severe aortic regurgitation ($P < 0.001$).

Postoperative and long-term results according to surgical procedure are summarized in Table 3 and Fig. 1 and are detailed in the following paragraphs.

Postoperative course

There were no operative deaths. We documented the following postoperative complications: low cardiac output [4 patients (13%) with 3 presenting as dilated cardiomyopathy at last follow-up]; pericardial effusion (3 patients, including 2 who required drainage); serious thoracic bleeding requiring reintervention in 1 patient; atrioventricular block (3 patients, including 2 who required a permanent pacemaker after a Bentall procedure); mediastinitis (1 patient); and transitory arrhythmia (6 patients). All the patients with postoperative low cardiac output had moderate or severe aortic regurgitation at surgery associated with a suspected or documented connective tissue disorder. The coronary arteries were assessed as normal by coronary angiography when it was performed.

Mortality rate

The median follow-up period in this study was 6.8 years (IQR 1.7–12.7 years). We reported 2 short-term cardiac-related deaths (6%) in patients with IMS who presented with severe left ventricular failure during their postoperative course or at a later point. One was a 5-year-old boy (aortic diameter 45 mm, *z*-score +8.6; bicuspid aortic valve) who had undergone a David procedure and mitral valve annuloplasty. The surgical result after implantation of a 26-mm aortic root graft was initially excellent, but 6 months later, he unexpectedly presented with dilated cardiomyopathy without identified cause. He died quickly despite medical treatment. The second patient was an 18-month-old girl who had undergone a Yacoub operation and an aortic valve repair because of moderate aortic regurgitation. The procedure in this case required a long cross-clamp time (222 min) due to the need for revision of the left coronary button anastomosis. The child left the operating room with severe cardiac dysfunction that never improved. She died 3 months later.

In addition, a 3-year-old patient with LDS died of a haemothorax probably related to malinsertion of a central line that was placed 2 months after the initial operation for antibiotic delivery.

The 1-, 5- and 10-year patient survival rate was 91%. By univariable analysis, patients who died of cardiac-related causes tended to have a higher preoperative aortic root *z*-score compared with survivors [+9.3 (IQR +9 to +9.7) vs +6.9 (IQR +5.5 to +7.8); $P = 0.07$; Table 4].

Table 3: Surgical outcomes according to the aortic root procedure

Surgical outcomes	Bentall ($n = 11$)	David ($n = 11$)	Yacoub ($n = 10$)
Postoperative complications, *n* (%)	4 (36)	3 (27)	3 (30)
Cardiac-related deaths, *n* (%)	0	1 (9)	1 (10)
Reoperation, *n* (%)	0	4 (36)	7 (70)
Follow-up (years)	7.7 (3–10.2)	6 (2.4–9.8)	13.5 (2.4–17.9)

Quantitative values are expressed as medians (interquartile ranges).

Postoperative complications include low cardiac output, mediastinitis, atrioventricular block requiring a permanent pacemaker, thoracic acute bleeding and pericardial effusion requiring drainage.

Reoperation

The median follow-up period for operative survivors ($n = 29$) was 7.7 years (IQR 4.2–12.8 years). Eleven patients (38%) required a reoperation after a median of 4.1 years (IQR 2–5 years) for significant aortic insufficiency despite a good immediate postoperative result (Table 5). In addition, 8 of them had an aneurysm of the aorta, distal to the aortic graft. All the children requiring a reoperation had initially undergone a surgical procedure with preservation of the native valve [Yacoub repair ($n = 7$) or David repair ($n = 4$)]. Therefore, at a median age of 9.8 years (range 6.1–12.3 years), these children needed an AAR with a mechanical prosthesis whose median size was 23 mm. There were no deaths or serious complications during the reoperations.

Three patients underwent a second reoperation: 2 required a mitral valve replacement with a mechanical prosthesis and 1 had a revision of a Bentall graft after 10 years for aortic stenosis.

Ten of the 11 patients who required reoperations (91%) had a diagnosis of IMS. Using a multivariable Cox model including IMS diagnosis and Yacoub procedure, it appeared that IMS condition tended to be more closely related to the risk of reintervention than the surgical procedure (Table 6). Freedom from aortic reintervention at 1, 5 and 10 years after AAR in IMS versus non-IMS children was, respectively, 100%, 33% and 25% versus 94%, 94% and 94% ($P = 0.02$; Fig. 2).

DISCUSSION

Timing for surgery

Because AAR is a prophylactic procedure performed to avoid life-threatening aortic aneurysm complications such as aortic dissection or rupture, the optimal surgical timing has to be particularly well managed. Although Roman *et al.* [9] have described the 'normal' range for aortic diameter in children (stratified by body surface area and age category) and Zanotti *et al.* [10] published guidelines for paediatric operative management regarding the underlying connective tissue disorder, the surgical decision-making algorithm remains controversial. Moreover, these guidelines might not be fully applicable in small children. Further, the specific criteria to use in children with IMS or LDS are

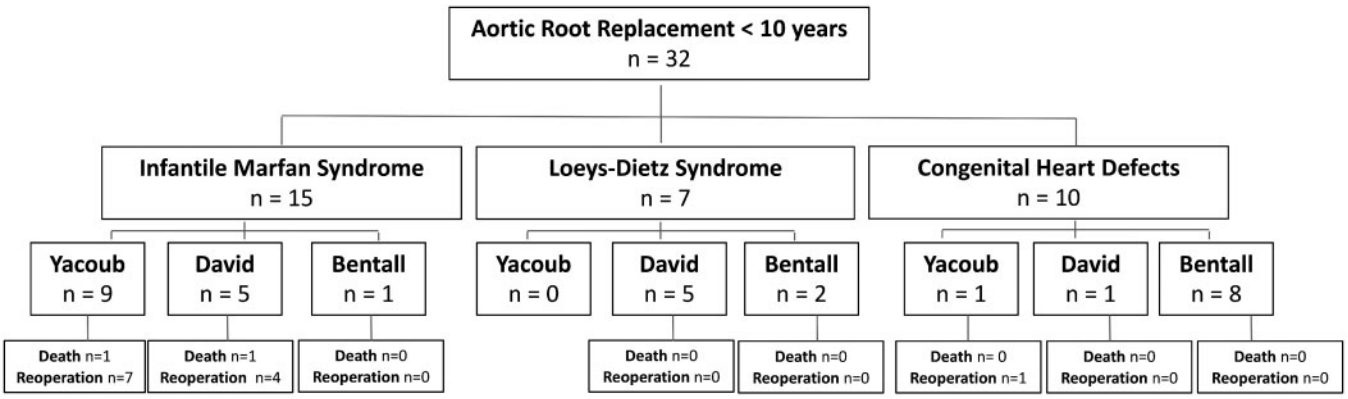

Figure 1: Flow chart illustrating the population included in the study and its outcome.

Table 4: Risk factors for cardiac-related mortality rates

Risk factors	Alive ($n = 29$)	Cardiac-related deaths ($n = 2$)	*P*-value
Male, *n* (%)	19 (66)	1	1
Age (years)	5.9 (3.8–7.6)	3.3 (2.4–4.1)	0.27
Weight (kg)	18 (13.3–24.5)	12.2 (10.7–13.6)	0.19
Preoperative aortic root *z*-score	+6.9 (+5.5 to +7.8)	+9.3 (+9 to +9.7)	0.07
IMS syndrome, *n* (%)	13 (45)	2	0.23
Connective tissue disease, *n* (%)	19 (66)	2	1
Aortic regurgitation, *n* (%)	12 (45)	1	1
Non-tricuspid aortic valve, *n* (%)	12 (41)	1	1
Previous operation, *n* (%)	9 (31)	1	1
Valve-sparing procedure, *n* (%)	19 (66)	2	1
Cross-clamp time (min)	108 (85–125)	169 (142–195)	0.15

Quantitative values are expressed as medians (interquartile ranges).
IMS: infantile Marfan syndrome.

questionable because the number of reported cases is limited and the risk of aortic dissection according to absolute diameters or *z*-score is unknown in this age group. Indeed, some authors apply the same criteria as that used in adults for prophylactic surgery [11], whereas others only consider the rate of progression of the diameter and the association with aortic regurgitation [12]. It should be noted, however, that the existing published data suggest that aortic root dilatation in children with IMS tends to progress with time causing a significant risk of dissection, rupture or death [13]. Regular surveillance for changes in aortic root diameter is necessary. We did not report any dissection in the paediatric age-group as an indication for surgery in the present cohort, though we did observe that perioperative death was significantly increased in patients with higher aortic root *z*-scores at the time of the operation. This increase in the number of deaths may be partly attributable directly to the severity of the underlying disease but also partly to the consequent technical difficulty of operating on a considerably enlarged aortic root. This situation suggests the potential utility of defining more precisely cut-off values for AAR in young children through the use of registry data or larger, multicentric longitudinal studies.

Table 5: Surgical and demographic features of the patients who required a reintervention with aortic valve replacement with a mechanical prosthesis

Patient	Connective tissue disorder	Age (years)	Operation	Aortic root graft #1 (mm)	Reoperative delay (years)	Aortic root graft #2 (mm)
1	IMS	0.8	Yacoub		11.1	24
2	IMS	4.8	Yacoub	20	5.0	24
3	IMS	4.6	Yacoub	14	2.0	22
4		9.3	Yacoub	20	0.7	22
5	IMS	2.8	Yacoub		4.4	28
6	IMS	6.2	Yacoub	18	4.0	24
7	IMS	3.4	Yacoub	16	5.0	24
8	IMS	5.0	David	20	4.1	22
9	IMS	4.5	David	22	1.6	24
10	IMS	4.2	David	19	8.1	28
11	IMS	9.7	David	24	2.1	28

IMS: infantile Marfan syndrome.

Table 6: Risk factors for reoperation (Cox proportional hazards analysis)

Variables	Hazard ratio	95% CI	*P*-value, univariable	*P*-value, multivariable
Male	1.7	0.46–6.7	0.42	
Age	0.91	0.72–1.1	0.41	
Weight	1	0.96–1.1	0.37	
Z-score	1.1	0.76–1.6	0.58	
IMS	12	1.6–97	0.017	0.042
Aortic regurgitation	0.28	0.061–1.3	0.11	
Yacoub procedure	4.2	1.2–4.6	0.022	0.74
Non-tricuspid aortic valve	0.8	0.21–3	0.74	

CI: confidence interval; IMS: infantile Marfan syndrome.

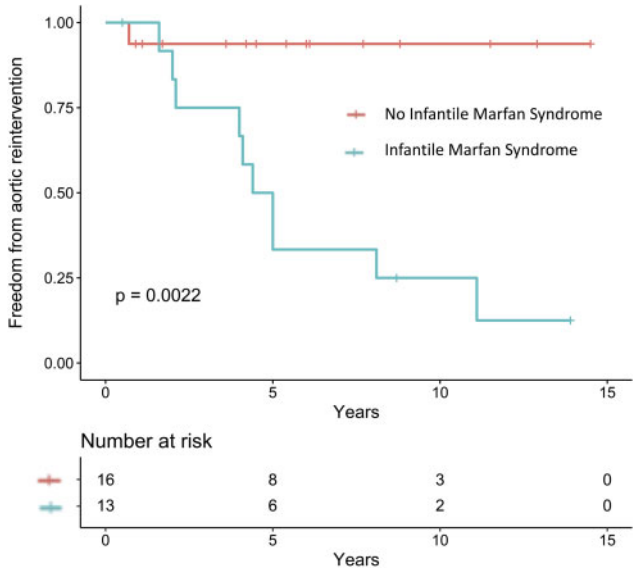

Figure 2: Kaplan–Meier graph showing freedom from aortic valve reintervention according to the underlying disease (infantile Marfan syndrome or not). Log-rank test: $P = 0.0022$.

Procedures

Valve-sparing procedures present a low operative risk and have already been established as a safe alternative to composite graft replacement when AAR is required in children [14].

Aortic regurgitation following valve-sparing procedures was the most common indication for reintervention and a matter of concern in the very sick cohort of young patients with IMS. This regurgitation may be the result of further annular dilatation occurring after a Yacoub procedure, though it may also occur due to distortion/damage of the valve itself or to reduction of the height of coaptation after repair or during follow-up. Several studies have shown the superiority of the reimplantation versus the remodelling procedure in terms of delaying an inevitable annular dilation and occurrence of aortic valve insufficiency [14]. It is probably important to prevent any annular diameter increase by employing a reimplantation technique or by adding an external ring support during the Yacoub procedure in cases where the size of the aortic annulus has reached an adult diameter. The use of a Dacron tube graft with preconfigured pseudosinuses (Gelweave^{TM} Valsalva graft, Terumo, Ann Arbor, MI, USA) has the advantage of stabilizing the aortic root while also mimicking the geometry of native aortic sinuses, leading to more favourable flow patterns and reduced shear stress on the cusps themselves [14].

If a Bentall procedure was performed as the first operation, no patients required a reintervention after a median follow-up of 7.7 years. The youngest patient was 7 months old and received a 19-mm mechanical prosthesis. Given the preoperative severe annular dilatation of these patients, implanting an adult-size substitute is technically less difficult than expected and compatible with good haemodynamic function during childhood. One patient developed endocarditis 3 years after AAR but did not require reintervention. No thromboembolic event was recorded. One patient with severe LDS suffered from retinal bleeds. Overall, the Bentall procedure appears to be a safe, low-risk operation in children, including from the perspective of potential anticoagulation-related morbidity.

Connective tissue disorders

MS is the most frequently occurring connective tissue disorder caused by a heritable genetic mutation [15, 16]. Cardiac operations, especially AAR, are well-documented in adults and teenagers with MS. The operative indications are standardized by the European and American Consortium [17, 18]. The long-term outcome of the various surgical techniques for aortic root replacement in these older patients has been widely described and does not show significant difference in terms of reintervention rates in a recent large meta-analysis [19]. However, specific information about the indications for and the outcomes of these procedures in the youngest children are lacking [20, 21].

Unlike classic MS, IMS cases present a severe phenotype from early childhood [22] due to an autosomal dominant variation in the *FBN1* gene, usually between exons 24 and 32 [23]. The prognosis of IMS is usually described as very poor and is related to the severity of rapidly progressive cardiovascular complications [24, 25]. Interestingly, our results suggest that early AAR is a reasonable and low-risk procedure for the treatment of IMS. Nevertheless, with or without annular reinforcement, recurrent or progressive regurgitation appears to affect the majority of patients with IMS. One hypothesis is that these aortic valves have already undergone important adverse structural changes due to the aggressive nature of the disease, leading to significant cusp elongation, increase in the length of the cusp free margins and decreased effective height of coaptation. Concerning the poor mid-term results in this specific young population of patients with IMS [26, 27], one might propose adopting a more aggressive approach towards the valve itself at the time of repair by systematically performing extensive resuspension of the cusps (through symmetrical plication in their mid-portions) to achieve a greater effective height of coaptation. This is similar to the parallel situation in adults, where it is mandatory to control the height of coaptation after repair. When patients needed a reoperation after an initial valve-sparing procedure, they consistently needed a larger prosthesis than that required at initial repair; in most cases, this was equivalent to a theoretical adult-sized prosthesis. Although many of the children with IMS in our study therefore required reoperation after the initial valve-sparing root replacement, we were able to achieve some beneficial effect in that the need for anticoagulation was avoided in the youngest patients and deferred for a period of time (until reoperation), thus potentially reducing the time-related cumulative incidence of morbidity related to IMS.

Interestingly, although LDS is also a severe aortic disorder that causes major aortic root aneurysms in childhood, the results of AAR using valve-sparing procedures are better than those in patients with IMS. No patients in our cohort required reoperation after a median follow-up of 4.5 years. Even patients with LDS are described as being at high risk of aortic dissection and rupture at a young age, arguing for an earlier prophylactic AAR [28]; patients with IMS and LDS in our series were operated on at comparable ages.

Limitations

The present study was a single-centre, retrospective report. Multivariable analysis of risk factors with various outcomes was not possible due to the small numbers of patients and outcome

events. Large multicentre studies would help to clarify the superiority of different treatment strategies. The heterogeneity of the underlying disease conditions in our patient cohort (including IMS, other genetic syndromes and complex CHD such as transposition of great artery or double outlet right ventricle) also limits the clinical inferences and conclusions that may be made. It should also be acknowledged that the group undergoing Bentall operations had neither an aortic root aneurysm nor connective tissue disorder.

CONCLUSION

In accordance with previous reports, we believe that the Bentall procedure is a safe and durable operation for aortic root aneurysms, even in very young patients. We further suggest that aortic valve-sparing procedures exhibit good long-term results except in children with IMS whose almost-inevitable aortic annular dilatation and aortic valve regurgitation mandate reintervention after a short period. Systematic annular support and aggressive aortic valve resuspension at initial valve-sparing root repair might be an option to enhance the limited durability of repair. A Bentall procedure might be considered in this subgroup of patients, regardless of aortic valve function, when AAR is required.

Conflict of interest: none declared.

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