RESEARCH

Surgical preference for aortic root and ascending aortic aneurysm in pediatric patients: single-center experience

Çocuk hastalarda aort kökü ve asendan aort anevrizmasında cerrahi tercihler: tek merkez deneyimi

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Abstract

Purpose: Aortic root and ascending aortic aneurysms are rarely encountered in pediatric patients. Our aim in this study was to compare our surgical approach and results in this patient group with similar studies in the literature.

Materials and Methods: This study retrospective in design and its cohort consists of pediatric patients who were operated on due to the diagnosis of aortic root and/or ascending aortic aneurysm. The etiologies of the patients that caused the aneurysm were investigated preoperatively, and the severity of the disease in the aortic valve and ascending aorta and surgical indications were revealed with detailed imaging methods. Operative data was then retrieved and early postoperative morbidity and mortality, as well as post-discharge aortic valve functions were evaluated. The obtained data was compared with those of similar articles in the literature.

Results: A total of eight pediatric patients were operated on with the diagnosis of aortic root and ascending aortic aneurysm. The average age of the patients was eleven (±4.03) years. The underlying cause of three (37.5%) patients was Marfan syndrome, two (25%) had bicuspid aortic valve and one (12.5%) had Wiscott-Aldrich Syndrome, previously operated sinus valsalva aneurysm and operated truncus arteriosus. There was 3rd degree aortic valve insufficiency in six patients (75%). The average aortic annulus diameter, mean sinus of valsalva diameter and ascending aorta diameter of the patients were 26.7 mm (±5.3), 40.9 mm (±9.7) and 37.8 mm (±9.2), respectively. The David I procedure was performed in three (37.5%) patients, while the Bentall procedure or aortic valve and ascending aorta replacement was performed on the other five patients. Non-cardiac mortality was observed in one (12.5%) patient, whereas native and mechanical valve dysfunction was not observed in any patient after discharge.

Öz

Amaç: Çocuk hastalarda aort kökü ve asendan aort anevrizmalarına nadiren rastlanmaktadır. Bu çalışmadaki amacımız, bu hasta grubundaki cerrahi yaklaşımımızı ve sonuçlarımızı literatürdeki benzer çalışmaları ile karşılaştırmaktır.

Gereç ve Yöntem: Bu çalışma, dizayn olarak retrospektif gözlemsel bir çalışma olup kohortunu aort kökü ve /veya asendan aort anevrizması nedeniyle opere edilmiş çocuk hastalar oluşturmaktadır. Hastaların preoperatif dönemde anevrizma sebepleri araştırılmış, detaylı görüntüleme yöntemleri kullanılarak aort kapak ve asendan aorttaki hastalığın ciddiyeti ve cerrahi endikasyonları ortaya konmuştur. Sonrasında, sırayla operasyona ait, erken postoperatif morbidite ve mortalite verileri toplanmıştır. Taburculuk sonrası aort kapak fonksiyonları takip edilmiştir. Elde edilen veriler literatürdeki benzer çalışmaların verileri ile kıyaslanmıştır.

Bulgular: Aort kök ve asendan aort anevrizması tanısı ile toplam 8 çocuk hasta opere edildi. Hastaların ortalama yaşı 11 (±4.03) idi. 3 (%37.5) hastanın altta yatan sebebi Marfan sendromu iken ,2 sinin (%25) biküspit aort kapağı, 1 er (%12.5) hastanın ise Wiscott-Aldrich Sendromu, öncesinde opere edilmiş sinüs valsalva anevrizması ve opere edilmiş trunkus arteriosusdu. 6 hastada (%75) 3. derece aort kapak yetmezliği mevcuttu. Hastaların ortalama aort anülüs çapı, sinüs valsalva ortalama çapı ve asendan aorta çapı sırasıyla 26.7mm (±5,3), 40.9 mm (±9,7) ve 37,8mm (±9,2) idi. 3 (%37.5) hastada David I prosedürü uygulanmışken, diğer 5 hastada Bentall prosedürü ya da aort kapak ve asendan aort replasmanı uygulanmıştır. 1 (%12.5) hastada non kardiyak mortalite izlenmiş olup, taburculuk sonrası takiplerde hiçbir hastada nativ veya mekanik kapak disfonksiyonu izlenmemiştir.

Conclusion: Both valve-sparing surgical procedures and other replacement techniques can be used safely in pediatric patients with aortic root and ascending aortic aneurysms.

Keywords:. Ascending aortic aneurysm, Pediatrics, Aortic valve

Sonuç: Hem kapak koruyucu cerrahi prosedürler, hem de diğer replasman teknikleri aort kök ve asendan aort anevrizmalı çocuk hastalarda güvenle kullanılabilmektedir.

Anahtar kelimeler: Asendan aort anevrizması, pediatrik, aort kapak

INTRODUCTION

Aortic root and ascending aortic aneurysms are rarely encountered in pediatric patients. Although these pathologies are frequently observed in children with connective tissue disease, they can also be seen in pediatric patients with bicuspid aortic valves, chromosomal anomalies (e.g., Turner syndrome, Wiskott-Aldrich syndrome) or conotruncal anomalies (tetralogy of Fallot, transposition of great arteries, truncus arteriosus, etc.)[1-3]. Infrequently, they can be seen sporadically in some pediatric patients[4].

Due to their potentially fatal complications (aortic dissection, aortic valve insufficiency, rupture), the diagnosis and treatment of this disease must be conducted effectively. Unlike adult patients, the type and timing of the surgical procedure to be performed in pediatric patients with aortic root and ascending aortic aneurysms, may vary depending on varied factors. In addition, a clear guideline for the approach to aortic aneurysms in pediatric patients has not yet been determined[1, 2].

Today, both valve-preserving surgical methods and mechanical valve replacement techniques are frequently used in pediatric patients with ascending aortic aneurysms[1]. Due to the need for lifelong anticoagulation, the risk of patient-prosthesis incompatibility, and the risks of bleeding and thromboembolism, valve-preserving surgical methods have begun to be applied with increasing frequency in pediatric patients[1,2]. However, in addition to the advantages of valve-sparing surgical techniques, they have their own limitations[3]. Some of these are the difficulties in application in bicuspid aortic valves, the possibility of recurrent aortic insufficiency that may develop in the aortic valve in the long term, and the risks of aneurysm development in unchanged aortic root tissues[3]. Due to all these controversial issues, the most appropriate surgical approach for children with ascending aortic aneurysms is still being investigated.

The hypothesis of our study is that both valve-sparing surgical approach and mechanical valve replacement methods can be successfully applied in pediatric patients with aortic aneurysm who have undergone replacement of the ascending aorta. In order to prove the hypothesis, we retrospectively examined the perioperative data of pediatric patients who were operated on in our clinic with a diagnosis of aortic root and ascending aortic aneurysm, and compared our clinical approach and results in this patient group with similar studies in the literature. The results obtained in this article, evaluated together with the results of similar articles, may provide additional contribution to surgical decision-making processes in this challenging patient group.

MATERIALS AND METHODS

Study population

The study was conducted in Ankara Bilkent City Hospital pediatric cardiovascular surgery department. The cohort of the study consisted of eight pediatric patients who were operated on for the diagnosis of aortic root and/or ascending aortic aneurysm between March 2019 and September 2023. The study was approved by Ankara Bilkent City Hospital Ethics Committee with approval number of E2-23-5648 dated 22.11.2023.

All pediatric patients under the age of 18 who applied to our department and were determined to undergo ascending aortic replacement by the pediatric cardiology and cardiovascular surgery council, as a result of a diagnosis of ascending aortic aneurysm and additional systemic diseases, were included in the study. Patients with saccular or fusiform aneurysms that developed in any segment of the aorta other than the ascending aorta, regardless of the cause, were excluded the study.

Preoperative imaging and surgical indications

Most patients were referred to our institution, which is one of the largest tertiary referral hospital of the country for surgery, after being diagnosed at another center. Echocardiographic studies of the hospitalized

patients were routinely performed and computed tomography scans were repeated if necessary. The underlying etiologies of the patients' aortic root and ascending aortic aneurysms were investigated and the surgical procedure to be applied (valve-sparing surgery (David), composite valve graft repair (Bentall procedure) or supracoronary graft interposition) was determined accordingly. The surgery indications in our clinic are as follows: 1) In patients with Marfan syndrome, the diameter of the aortic root or ascending aorta should be over 5 cm, or if there is accompanying significant aortic insufficiency, it should be over 4.5 cm. 2) In patients with aortic aneurysm with bicuspid aortic valve, the aortic diameter (root or ascending aorta) should be over 5.5 cm or if there is an indication for valve intervention, it should be over 4.5 cm. 3) In aortic root aneurysm due to other chromosomal anomalies, previous conotruncal operation or Ross surgery, the diameter should be over 5.5 cm. 4) Aortic enlargement should be more than 0.5 cm/year, regardless of the type of pathologies. Regardless of the patients' diagnoses, the operations were performed by three separate senior cardiac surgeons experienced in both valve-sparing aortic surgery and mechanical aortic valve replacement. Different strategies have been applied in surgical intervention and timing depending on the morphology and insufficiency of the aortic valve. The preferred surgical method in patients with Marfan and Wiskott-Aldrich syndromes was valve-sparing surgery. In patients with bicuspid valve, with significant valve insufficiency where repair is not feasible, the aortic valve is replaced with a mechanical valve. However, the final decision regarding the use of valve –sparing technique or composite valve graft repair in these cases was left to discretion of the senior surgeon who performed the operation.

Postoperative management

Echocardiographic studies of the operated patients were re-performed before discharge. Warfarin treatment was started in patients who underwent composite mechanical valve conduit repair, aiming for an INR value between 2.0-2.5. In patients who underwent valve-sparing aortic root or ascending aortic replacement, acetylsalicylic acid treatment at a dose of 5 mg/kg (maximum 100 mg/day) for three to six months was prescribed.

After discharge, patients were routinely followed up by pediatric cardiology in the 1st, 3rd, 6th, 12th months and then annually. During the follow-ups, particular attention was paid to the severity of aortic valve insufficiency and left ventricular functions.

Operative technique

A total of three patients with Wiscott-Aldrich and Marfan syndrome were successfully operated on using the valve-sparing David I reimplantation procedure, as described in the literature[5]. We determined the tubular graft size used in the David I procedure by measuring the diameter of the imaginary sinotubular junction, which is obtained by hanging the aortic commissures symmetrically in the superior direction, as described by Khachatryan et al.[6]. Aortic root and arch aneurysm had developed in a patient who previously underwent an isolated supracoronary graft interposition, due to the Marfan syndrome. In this patient, the Bentall procedure combined with total arch replacement was performed instead of David I, because the surgical procedure was overly complicated and the adhesions did not allow adequate tissue dissection. In the intraoperative evaluation of patients with bicuspid aortic valves, it was determined that the valves had thickened and begun to calcify, so aortic valve replacement combined with supracoronary graft interposition was performed with the decision of the primary surgeon. Similarly, aortic valve replacement, supracoronary graft interposition and replacement of the valved conduit in the right ventricular outflow tract, were performed in a patient who had previously undergone total correction of the truncus arteriosus. Severe aortic regurgitation and ascending aortic pseudoaneurysm developed secondary to infective endocarditis in a patient who had previously undergone repair for a sinus valsalva aneurysm. In this patient, aortic root and ascending aorta replacement was performed using the Bentall procedure. Antegrade selective cerebral perfusion technique was used in patients who underwent total arch replacement and ascending aortic pseudoaneurysm repair.

Statistical analysis

Categorical measurements were summarized as numbers and percentages, and numerical measurements were summarized as mean and standard deviation. All statistical analyses were performed using IBM SPSS (Statistical Package for the Social Sciences) Statistics ver.25.

RESULTS

The average age and weight of the eight patients included in the study was 11.0 (±4.03) years and 39.6 (±28.4) kg, respectively. Seven of the patients were male and one was female (85.5%-14.5%). The underlying cause of aneurysm in three (37.5%) patients was Marfan syndrome, while two (25%) had

bicuspid aortic valve. One (12.5%) patient had Wiscott-Aldrich Syndrome, previously operated sinus valsalva aneurysm and previously operated truncus arteriosus. In five patients (62.5%), there was an additional comorbidity factor. Reoperation was performed in three patients (37.5%). The demographic and clinical characteristics of the patients are shown in Table 1.

Table 1. Demographic and clinical characteristics of the patients

Patient #	Age(year)	Gender	Weight (kg)	Underlying Pathology	Additional Comorbidity
1.	9	Male	20	Wiscott-Aldrich Syndrome	Bone Marrow Transplantation , Previous Cerebrovascular Accident
2.	3,5	Female	18	Marfan Syndrome	Pectus Carinatum
3.	16	Male	105	Marfan Syndrome	Bilateral Lens Subluxation
4.	15	Male	45	Bicuspid Aortic Valve	Pulmonary Hypertension
5.	14	Male	45	Bicuspid Aortic Valve	Previous Stent implantation for Aortic Coarctation and Cerebrovascular Accident(right hemiplegia)
6.	9	Male	21	Truncus Arteriosus(Operated)	RV-PA Conduit obstruction
7.	11	Male	25	Sinus Valsalva Aneurysm(Operated)	Infective Endocarditis and Ascending Aorta Pseudoaneurysm
8.	11	Male	34	Marfan Syndrome	Operated Ascending Aortic Aneurysm (with Dacron Conduit) and Aortic Valve Repair
Mean (Standard Deviation)	11(±4)	-	39,6(±28,4)	-	-

RV; Right ventricle, PA; Pulmonary artery

Preoperative echocardiographic findings of the patients are presented in Table 2. The ejection fraction of all patients was above 60%. Aortic valve stenosis was not observed in any of the patients. While six patients (75%) had 3rd degree aortic valve insufficiency, the aortic valve in six patients (75%) had tricuspid valve morphology. While the average aortic annulus diameter of the patients was 26.7 mm (±5.3), the patient with the narrowest annulus had an annulus of 19 mm. The mean annulus Z score of the patients was 5,0 (±2,9). The mean sinus valsalva diameter and Z scores of the patients were 42,3 (±9,9) mm and 6,6 (±3,5), respectively. The patients' ascending aortic diameter and Z scores were 37,8 (±9,2) mm and 6,9 (±2,5), respectively.

The perioperative data is presented in Table 3. The David I procedure was performed in three (37.5%) patients, while the Bentall procedure or aortic valve and ascending aorta replacement was performed in the other five (62.5%) patients. The aortic annulus diameters of all operated patients were sufficient for both mechanical valve implantation and valve-sparing surgical procedures (26.7mm ± 5.3). In this way, neither the need for root expansion nor patient-prosthesis mismatch was observed in any patient. A 28- or 30-mm Dacron tube graft could be implanted into the ascending aorta in all patients. In patients with bicuspid aortic valve and in the patient with aortic root aneurysm following truncus arteriosus repair, mechanical valve and ascending aortic replacement was performed without intervening the sinus valsalvae, with the decision of the primary

surgeon. The surgical procedures were successfully performed on all patients. The mean cardiopulmonary bypass (CPB) time was 194.75 (±47.3) minutes, the mean cross-clamp time was 139 (±30) minutes, and the mean antegrade selective cerebral perfusion (ASCP) time was 17 (±5.5) minutes. No hemodynamic instability was observed in the follow-ups of patients during postoperative intensive care unit period.

Pleural and pericardial effusion requiring intervention developed in one patient each. Left bundle branch block was observed in one patient. While no mortality was observed due to cardiovascular reasons in any operated patient, one patient (12.5%) died during the in-hospital period due to acute intracranial hemorrhage. All other patients (87.5%) were discharged under close follow-up, with no cardiovascular problems.

Table 2. Preoperative echocardiographic parameters of the patients

No	Underlying Disease	EF (%)	AV Ana.	AV Reg.	AA (mm)	AA Z skor	SV (mm)	SV Z score	AsA (mm)	AsA Z Score
1.	Wiscott-Aldrich Syndrome	65	TC	3	29	7,52	46	9,59	42	9,78
2.	Marfan Syndrome	48	TC	3	37	10,26	45,7	9,69	34,5	7,99
3.	Marfan Syndrome	70	TC	Trivial	28	2,30	45	3,94	38	3,7
4.	Bicuspid Aortic Valve	79	BC	3	22	2,38	35	3,91	37	6,0
5.	Bicuspid Aortic Valve	59	BC	3	25	3,65	53	8,52	58	10,5
6.	Truncus Arteriosus (Operated)	68	TC	3	28	7,02	48	9,9	33	7,22
7.	Sinus Valsalva Aneurysm (Operated)	70	TC	1	19	2,63	21	0,15	33,5	6,69
8.	Marfan Syndrome (Operated)	62	TC	3	26	4,87	45	7,62	27	3,74
Mean (Standard Deviation)		65 (±9)			26,7 (±5,3)	5,0 (±2,9)	42,3 (±9,9)	6,6 (±3,5)	37,8 (±9,2)	6,9 (±2,5)

EF;Ejection Fraction ,AV Ana; aortic valve anatomy, , AV Reg; Aortic valve Regurgitation, AA; aortic annulus, SV; sinus valsalva, AsA; Ascending AortaTC; tricuspid ,BC; Bicuspit

In patients who had 3rd degree aortic valve insufficiency in the preoperative period and underwent valve-sparing reimplantation technique, trivial or 1st degree insufficiency was observed in the early postoperative period. No significant valve dysfunction or patient-prosthesis mismatch was detected in patients who underwent mechanical aortic valve replacement. During an average follow-up of 20.6 (±9.5) months, no increase in aortic valve insufficiency was observed in any of the three patients who underwent valve-sparing procedure. No thromboembolic events or bleeding complications were observed in any patient discharged during this period. Parameters following operation and discharge are presented in Table 4.

Table 3. Perioperative data

No	Underlying Pathology	Operation	CPB (min)	Cross Clamp (min)	ASCP (min)	ICU Stay (day)	Total Hospital Stay (day)	In Hospital Mortality	Postoperative Complication
1.	Wiscott-Aldrich Syndrome	David (30 mm Dacron)	180	135	0	2	6	No	Massive Pleural Effusion
2.	Marfan Syndrome	David (30 mm Dacron)	187	132	0	2	11	No	-
3.	Marfan Syndrome	David (30 mm Dacron)	172	142	0	1	6	No	Massive Pericardiac Effusion
4.	Bicuspid Aortic Valve	21 No SJM AVR+ Supracoronary Graft (30 mm Dacron)	176	148	0	1	8	No	Left Bundle Branch Block
5.	Bicuspid Aortic Valve	25 No SJM AVR + Supracoronary Graft (30 mm Dacron)	170	136	0	15	16	**Yes**	Massive Intracranial Hemorrhage
6.	Truncus Arteriosus (operated)	23 No SJM AVR+ Supracoronary Graft (28mm)+ RV-PA Conduit Replacement	187	105	0	3	12	No	-
7.	Sinus Valsalva Aneurysm (operated)	Bentall Procedure (21 No SJM) + Aortic Pseudoaneurysm Repair	311	206	13	6	21	No	-
8.	Marfan Syndrome (Operated)	Bentall Procedure (25 No SJM)+ Total Arc Replacement (30mm)Dacron	175	115	21	2	14	No	-
Mean (Standard Deviation)		-	194(± 47,3)	139(± 30)	17(5,6)	4(±4, 7)	11,7(5,2)	-	-

CPB; Cardiopulmonary bypass, ASCP; antegrade selective serebral perfusion, ICU; intensive care unit,SJM; St. Jude Medical

Table 4. Parameters following operation and discharge

No	Underlying Pathology	Operation	Preoperative AV Regurgitation	Early Postoperative AV Regurgitation	Average Postoperative Follow-up Time (months)	AV regurgitation following discharge	Thromboembolic event or bleeding following discharge
1.	Wiscott-Aldrich Syndrome	David (30 mm Dacron)	3	1	30	1	No
2.	Marfan Syndrome	David (30 mm Dacron)	3	Trivial	24	Trivial	No
3.	Marfan Syndrome	David (30 mm Dacron)	Trivial	Trivial	25	Trivial	No
4.	Bicuspid Aortic Valve	21 No SJM AVR+ Supracoronary Graft (30 mm Dacron)	3	Trivial	24	Trivial	No
6.	Truncus Arteriosus (operated)	23 No SJM AVR+ Supracoronary Graft (28mm)+ RV-PA Conduit Replacement	3	Trivial	26	Trivial	No
7.	Sinus Valsalva Aneurysm (operated)	Bentall Procedure (21 No SJM) + Aortic Pseudoaneurysm Repair	1(paravalvular)	Trivial	5	Trivial	No
8.	Marfan Syndrome	Bentall Procedure (25 No SJM)+ Total Arc Replacement (30mm)Dacron	3	Trivial	6	Trivial	No
	Mean (Standard Deviation)	-	-	-	20,6(±9,5)	-	-

AV; Aortic valve , SJM; St. Jude Medical, AVR; Aortic valve replacement

DISCUSSION

The incidence of aortic root and ascending aorta aneurysm in pediatric patients is not patently known. However, in autopsy studies conducted following sudden cardiac deaths in young people, aortic aneurysms were found in 5.4% of the patients[7]. Connective tissue diseases are the most common cause of aneurysms requiring surgery in pediatric patients, with a rate of 90%[2]. Apart from these, some chromosomal anomalies (Down syndrome, Turner syndrome, etc.), the presence of bicuspid aorta or conotruncal anomaly, previous pulmonary autograft implantation (Ross procedure), some pathologies with single ventricle physiology, aortic coarctation, aortitis and some autoimmune vasculitis, may cause aneurysm development in the aorta[7]. However, idiopathic cases unrelated to any factor have also been reported[8].

Among connective tissue diseases causing aortic aneurysms, the Marfan and Loeys–Dietz syndromes are the most common. Both diseases have autosomal dominant inheritance and are associated with aortic aneurysms, that develop secondary to the deterioration of the structural resistance of the aortic media layer due to genetic mutations. The aneurysms observed in these patients can sometimes expand very rapidly and lead to the death of the patient with catastrophic consequences (such as aortic dissection, rupture). However, there may be clinical variability in each disease due to differences in genetic penetrance or phenotypic features according to age[9]. Therefore, close cardiological follow-up of patients in both groups is essential.

Many pathologies other than connective tissue diseases can cause aortic root and ascending aortic aneurysm in the pediatric age group. Among these,

having a bicuspid aortic valve is one of the leading congenital causes, with a rate of 2% in the population. In these patients, where the aortic valve functions may be normal[4], medial degenerative changes occurring in the ascending aortic walls may cause aortic dilatation in approximately 50% of male patients. Unlike Marfan syndrome, while the aortic root is preserved in most patients, the aneurysm often manifests itself between the sinotubular junction and the innominate artery. However, in other, less common phenotypes, the entire aortic root and ascending aorta may be diffusely involved. Postmortem studies have found that the risk of aortic dissection is eighteen times higher in patients with bicuspid aortic valves than in those with tricuspid valves[9].

Apart from these, we currently encounter patients who were previously operated on for congenital heart disease and developed aortic root aneurysm during follow-up. These patients were operated on due to conotruncal anomalies (tetralogy of Fallot, truncus arteriosus, d-TGA, etc.), or patients who have undergone Ross surgery or single ventricle palliation. There remains uncertainty in the literature regarding the guideline to be followed in this patient group. However, the limited experience gained to date indicates that the likelihood of encountering catastrophic complications in the early period due to aortic root aneurysm, is low in these patient groups[1].

The issue of optimal surgical timing in pediatric patients with aortic aneurysm is still controversial and varies depending on the type and phenotypic severity of the underlying disease. Therefore, the typical approach is to closely follow all patients with dilatation and determine the surgical strategy according to patient's own characteristics. Accordingly, in patients with Marfan syndrome, it is recommended to wait until aortic root dilatation is 5 cm before surgical intervention, as dissection or rupture is rarely observed in early childhood. However, if there is an annual growth of more than 0.5 cm or if there is a family history of early rupture, surgery can be performed at an earlier period. Since aortic dissection and rupture can be observed in early childhood in the Loeys–Dietz syndrome Types 1 and 2, aortic diameter > 3.5 cm or Z score > 3 and annual growth amount more than 0.5 cm have been determined as surgical indications. However, in this group, in order to prevent prosthesis-patient mismatch in the future, close follow-up with the most intense medical treatment possible is recommended

to ensure that the aortic annulus diameter of the patients reaches at least 18 mm[3]. Today, the lower limit of surgical intervention in operated Ross patients who develop aortic aneurysm is accepted as 5 cm. On the other hand, in aortic aneurysms that develop after operated conotruncal anomalies, aortic coarctation and single ventricle palliations, if there is no aortic valve insufficiency, it is recommended to wait for 5.5 cm for aortic root intervention[1,2]. In adults, there are publications attempting to predict aortic complications by indexing the aortic cross-sectional area to the patient's body surface area or height[10,11]. Although successful results have been achieved in risk stratification in adult patients, these indices have not yet been sufficiently validated in pediatric patients. Therefore, the question of when to consider surgical treatment in pediatric patients, based on which patient, at what aortic diameter, as well as with which clinical and radiological findings, remains unclear. It is evident that large-scale randomized controlled trials are needed in this regard[1].

To date, a wide variety of surgical strategies have been described in patients with aortic root and ascending aortic aneurysms. Robichec et al.[12] suggested the wrapping of ascending aorta with artificial vascular graft after elliptical excision. Sarsam et al.[13] described the valve-sparing remodelling technique, in which the aortic root cannot be completely stabilized. Recently, the Florida Sleeve technique, in which the aortic root is stabilized and sinus valsalva is preserved by wrapping the aorta entirely, has been described[14]. Lately, the PEARS (personalized external aortic root support) technique, which can be considered as a different modification of original wrapping technique, has been introduced[15]. However, there are a few techniques that can be used safely in pediatric patients and whose long-term results are well-known. Among these, the most frequently used and popular one is the aortic valve-sparing reimplantation technique[5,16]. Thanks to this technique, which was first described by Tirone David and has undergone many modifications over the years, the ventriculoaortic junction can be safely stabilized and the entire valvular apparatus can be protected. Having completely resected of the sinus valsalva and the ascending aorta, there is no residual aortic tissue left that can cause recurrent aneurysm with this technique. After replacing the entire root and ascending aorta using Dacron tube grafts (Valsalva or straight grafts), the native aortic valve and coronary arteries can be reimplanted into the tube graft. In the

study conducted by Fraser et al., which included 100 pediatric patients who underwent valve-sparing surgery, early mortality was observed in only two of the patients (2%). The late reintervention-free rate of this technique was found to be 94.5% in five years and 78.6% in ten years. During an average of seven years of follow-up, the need for aortic valve replacement was reported to be approximately 6%. Additionally, only four out of 84 valve-sparing surgical procedures (4.8%) showed annular enlargement and severe aortic valve insufficiency, which may require AVR in the late period[2]. When all these results are evaluated, it can asserted that the valve-sparing reimplantation technique has successful results and can be safely applied in every anatomically appropriate pediatric patient. In our own clinic, we performed valve-sparing root and ascending aortic replacement in a total of three patients with Marfan and Wiskott-Aldrich syndromes. We did not encounter any complications in the intraoperative or early postoperative period in any of these three patients. We did not observe any increase in aortic valve insufficiency, decrease in valve movements or coaptation defects in any of our patients, during follow-up. We continue to follow all our patients closely for the progression of aortic valve insufficiency, as it has been reported in the literature in the long term.

Despite satisfactory mid- and long-term results[2,17], careful selection of patients who will undergo valve-sparing reimplantation technique is essential to prevent postoperative mortality and reintervention. This is especially important in patients with bicuspid aortic valves. Preserving the bicuspid valve either in its native form or carefully determining the location of the commissural crests within the graft during the reimplantation phase after its repair, is essential to prevent aortic valve insufficiency after implantation. Aicher et al. reported very successful results in terms of being free from aortic valve insufficiency in the medium and long term, in patients on whom they performed bicuspid valve repair in addition to valve-sparing root replacement[18]. Similarly, Fraser et al. reported that in the long-term follow-up of twelve pediatric patients with bicuspid valves, in whom they successfully performed aortic root replacement with valve-sparing techniques, none of them required reoperation due to severe aortic regurgitation[2]. Although the presence of a bicuspid valve itself does not constitute a contraindication, it is recommended that valve-sparing surgery should not be performed, especially in patients with valve calcification, stress-related asymmetric prolapse and commissural fenestrations, or immobile valve movements that cannot be repaired. In addition, excessive fusion between the aortic wall and myocardium (ventricularization) that prevents their separation from each other is also considered a relative contraindication for valve-sparing surgery[2]. In these patients, intolerable aortic valve insufficiency may be observed after the repair procedure. Therefore, replacement with a mechanical valve may be a valid alternative in patients whose aortic valve is affected by the underlying disease and in bicuspid aortic valves that are not suitable for repair. Replacement was applied to two patients with bicuspid valves in our study group because their valve anatomy was not suitable for repair. Postoperative follow-up of these patients was uneventful.

Ascending aorta replacement performed using a composite valve graft conduit is a successful method with well-known long-term results in the adult patient group. The results of this surgical method in pediatric patients are also satisfactory[19,20]. Studies have reported a 10-year survival rate of 100%[21]. However, the fact that this surgery has some disadvantages (the need for lifelong anticoagulation, the possibility of patient-prosthesis incompatibility, the risk of thromboembolic events or bleeding) has caused surgeons to consider different surgical alternatives in pediatric patients. In cases where no valve repair method can be feasible and in hemodynamically urgent cases, such as acute dissection or rupture, the Bentall procedure can still be used if the annulus diameter is appropriate (at least 18 mm)[22,23]. Long-term studies have shown that the incidence of thromboembolic events is lower in pediatric patients compared to adults[23]. In pediatric patients where anticoagulation is contraindicated, xenograft composite conduits with or without stents can also be used, although their long-term results are unsuccessful. Since all patients in our sample had an aortic annulus over 18 mm, we did not encounter any difficulties in either replacement or valve-sparing procedures. All our patients to whom we placed a composite mechanical valve conduit were effectively anticoagulated with warfarin treatment, and no thromboembolic or bleeding requiring intervention was encountered during an average follow-up period of 20.5 (±9.5) months.

Although not frequently preferred today, aortic homografts can also be used in young children with narrow annulus[22]. However, it is known that these

grafts do not last long in pediatric patients and half of the patients must be re-operated within two to five years due to graft dysfunction, which pose serious operative risks secondary to severe reactions and calcification in these patients. Therefore, most clinics prefer to use aortic homografts only if a suitable prosthetic valve cannot be implanted. The typical approach in this patient group is to monitor the patients closely and let them grow with medical treatment until they are suitable for valve-sparing surgery or the Bental procedure (at least 18 mm). Thus far, no patient with an annulus size below 19 mm has been admitted to our clinic. However, in case of such a situation, we plan to follow-up these patients with medical treatment instead of urgent surgery, as recommended in the literature.

Despite successful aortic root and ascending aortic replacement, recurrent aortic dissection or aortic arch aneurysm may develop in the long term, especially in pediatric patients with connective tissue disease. It is still controversial to replace all residual aortic segments, which may be structurally deteriorated in later ages, although they have normal diameters and structure in the first operation. In the study conducted by Fraser et al., only one out of ninety patients with Marfan and Loeys–Dietz syndromes underwent arch intervention in the first surgery. In eight of the remaining patients, the need for reintervention in the aortic arch occurred after an average of seven years of follow-up. Six of these patients had Loeys–Dietz subtypes with an aggressive course[2].

In Loeys–Dietz syndrome in particular, because the aortic structures grow very rapidly and are prone to rupture, some clinics advocate the necessity of a major intervention in the aortic arch in this patient group, even if its dimensions are within normal limits in the first surgical operation. However, other groups state that the approach on this issue can be made on a patient and disease-specific basis and that arch reconstruction can be limited in patients with thin-walled, fragile and dilated arch tissue[4]. In our clinic, we performed isolated ascending aorta replacement without prophylactic arch reconstruction, in patients with Marfan syndrome who have normal aortic arc due to the risk of perioperative morbidity. Based on the late-term complications described in the literature, we follow up all our patients with close imaging (Echocardiography and CT) and monitor them for possible recurrent aneurysmal formations.

The small number of cases is the most obvious limiting factor of this article. Furthermore, due to its retrospective and observational design, it retains the inherent limitations of such studies. The absence of a prospective comparative analysis between valve preservation and mechanical valve replacement techniques in aortic aneurysms stemming from the same underlying active pathology, hinders the determination of the most effective and sustainable treatment approach for this patient cohort.

In conclusion, aortic root and ascending aortic aneurysms are rarely encountered in pediatric patients. Although most of them are due to connective tissue diseases, these aneurysms may occur secondary to some existing congenital cardiac diseases or previous cardiac surgery. Based on the results in the literature and our clinical experience, we believe that both valve-sparing surgical procedures and other replacement techniques can be used safely in pediatric patients with aortic aneurysm, resulting in low mortality and morbidity. Large-scale and multicenter prospective controlled studies to be conducted in the future may enable the determination of the most effective and reliable surgical method in the long term in this patient group.

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