BRIEF ARTICLE

Valve-Sparing Aortic Root Replacement in Children and Adolescents: Experience and Results in a Public Institution in Argentina

Reemplazo de la raíz aórtica con preservación valvular en niños y adolescentes: experiencia y resultados en una institución pública en Argentina

SANDRA SEPÚLVEDA[1], JUAN TORRILLAS[2], ANALÍA MARTIN[1], JULIANA MEDINA[1], JULIA BLANDO[1], GUILLERMO MORENO[3], MARIELA MOURATIAN[1], PABLO GARCÍA DELUCIS[2], GLADYS SALGADO[1, MTSAC]

ABSTRACT

Background: Valve-sparing aortic root replacement (VSARR) with reimplantation technique is indicated in patients with aortic root aneurysm.
Objective: To evaluate the mid-term outcomes of this surgical technique in children and adolescents in our institution.
Methods: Retrospective study. Twenty-one patients who underwent VSARR surgery were included between July 2006 and July 2022. The relationship between baseline variables and progression of aortic regurgitation (AR) was assessed.
Results: The median age was 13 years. Seventeen patients had connective tissue disorders (3 with bicuspid aortic valve), 2 operated congenital heart disease, 1 isolated bicuspid aortic valve and 1 Turner syndrome. Thirteen patients had mild AR and 5 had moderate AR preoperatively.
Three patients underwent early aortic replacement, 2 due to endocarditis and 1 due to severe acute AR. The median follow-up was 4.1 years. Of 17 patients at follow-up, 2 developed mild-moderate AR, 2 moderate AR and 3 severe AR. One of them underwent Bentall surgery. No association was found between the degree of preoperative AR or the presence of bicuspid aortic valve and the progression of postoperative AR. There was no association between the type of postoperative valve coaptation and the development of AR greater than mild at follow-up.
Conclusions: VSARR with reimplantation is a feasible surgical technique with favorable mid-term outcomes and low reoperation rate in pediatric patients.

Key words: A root aneurysm - Aortic root replacement - Valve-sparing.

RESUMEN

Introducción: La cirugía de reemplazo de la raíz aórtica con preservación valvar tipo reimplante (RRAoPV) está indicada en pacientes con aneurisma de la raíz aórtica.
Objetivo: Evaluar los resultados a mediano plazo de esta técnica quirúrgica en niños y adolescentes de nuestra institución.
Material y métodos: Estudio retrospectivo. Se incluyó a 21 pacientes a quienes se realizó cirugía de RRAoPV entre julio de 2006 y julio de 2022. Se evaluó la relación entre las variables basales y la progresión de insuficiencia aórtica (IAo).
Resultados: La mediana de edad fue de 13 años. Diecisiete pacientes tenían enfermedad del tejido conectivo, (3 de ellos además con aorta bicúspide), 2 cardiopatías congénitas operadas aisladas, 1 válvula aórtica bicúspide aislada y 1 síndrome de Turner. Trece pacientes tenían IAo leve y 5 moderada en el preoperatorio.
Tres pacientes requirieron reemplazo aórtico precoz, 2 por endocarditis y uno por IAo aguda grave. La mediana de seguimiento fue de 4,1 años. De 17 pacientes en seguimiento, 2 evolucionaron con IAo leve-moderada, 2 moderada y 3 grave. Uno de ellos requirió cirugía de Bentall. No se encontró asociación del grado de IAo preoperatoria, ni de la presencia de aorta bicúspide, con la progresión de la IAo postoperatoria. No hubo asociación entre el tipo de coaptación valvular postoperatorio y el desarrollo de la IAo mayor que leve en el seguimiento.
Conclusiones: El RRAoPV tipo reimplante es una técnica quirúrgica viable en pacientes pediátricos con buenos resultados a mediano plazo y baja tasa de reoperación.

Palabras clave: Aneurisma raíz aórtico - Reemplazo raíz aórtica - Preservación valvular.

Rev Argent Cardiol 2024;92:217-221. http://dx.doi.org/10.7775/rac.v92.i3.20768

Received: 11/27/2023 – *Accepted:* 03/24/2024

Correspondence: Sandra Sepúlveda. Combate de los Pozos 1801 (1245). E-mail: sandrasepulveda21@gmail.com

[1] Cardiology Service. Hospital de Pediatría "Dr. Juan P. Garrahan". Buenos Aires, Argentina.
[2] Cardiovascular Surgery Service. Hospital de Pediatría "Dr. Juan P. Garrahan". Buenos Aires
[3] Cardiovascular Recovery Service. Hospital de Pediatría "Dr. Juan P. Garrahan". Buenos Aires

INTRODUCTION

Aortic root aneurysms are rare in children and are generally associated with connective tissue disorders, (1,2) such as Marfan or Loeys-Dietz syndromes, or with the progression of conotruncal heart diseases. (2) These aneurysms may result in aortic rupture, aortic dissection or severe aortic regurgitation (AR). (2,3) Aortic root replacement is indicated to avoid the risk of these complications. Clinical guidelines and surgical indication are based on aortic root size, progression of dilatation (mm/year) and genetic syndrome.

The purpose of Tirone David surgery is to replace the pathological segment of the aortic (Ao) root complex while preserving the anatomical and functional integrity of the valve leaflets. (4) The advantage over Bentall surgery (aortic root replacement with a mechanically valved conduit) is that it avoids lifelong anticoagulation and its inherent risks. However, the main limitation of valve-sparing aortic root replacement (VSARR) in pediatric population is the potential need for reoperation, as during growth, the reconstructed aortic root and the aortic valve may become small for the future body surface area. (4)

This study evaluates our experience and the outcomes of VSARR with the Tirone David technique at the Hospital Nacional de Pediatría J.P. Garrahan.

METHODS

A retrospective study was performed. The medical records and echocardiographic recordings of 21 patients who underwent VSARR with the Tirone David technique between July 2006 and July 2022 were reviewed. The degree of AR, the valve morphology, the aortic root measurement and the Z-scores were assessed by transthoracic echocardiography and intraoperative transesophageal echocardiography (iTEE). Another imaging studies (cardiac tomography or resonance) were performed in 60% of patients.

Statistical data were analyzed with the STATA software. Continuous variables are expressed as median and interquartile range (IQR). Categorical variables are expressed as frequencies and percentages. The relationship between the basal variables and the progression of AR was evaluated.

RESULTS

Preoperative characteristics

The median age at surgery was 13.1 (10.5-14.4) years; the median weight was 40 (13-99) kg. The youngest patient was 2.9 years old and weighed 13 kg. Five patients were younger than 8 years. Eleven patients were female. All surgeries were scheduled and indicated based on the aortic root size and progression of aortic root dilatation (according to clinical guidelines). Seventeen patients had a diagnosis of connective tissue disease: 13 Marfan (2 associated with bicuspid aortic valve disease), 3 Loeys-Dietz (one associated with conotruncal heart defect) and 1 uncharacterized; 1 patient had Turner syndrome with bicuspid aortic valve disease; 2 isolated operated congenital heart diseases (1 conotruncal heart defect and 1 multiple ventricular septal defect cerclage), and 1 patient isolated bicuspid aortic valve disease.

The median diameter of the aortic annulus was 25 mm with a Z-score of +3.5 (+2.85-+5.7), and that of the Valsalva sinuses was 42.5 (36.5-51.5) mm with a Z-score of +6.2 (+4.6-+7.5). Regarding preoperative AR, 2 patients had no AR, 13 patients had mild AR, 1 mild-moderate AR and 5 patients had moderate AR.

Surgical data

The Tirone David technique was used in all patients. The smallest conduit size was 22 mm and the largest was 30 mm. All patients had a straight conduit. The median time of extracorporeal circulation (ECC) was 179 min and that of clamping 156 min. The longest ECC times were a consequence of associated procedures (pulmonary homograft replacement, mitral plastic surgery, transverse arch and descending aorta replacement). Only one patient (with Marfan syndrome and bicuspid aortic valve) was reoperated during the same procedure to resuspend the valve.

The iTEE performed in all patients showed trivial AR in fifteen patients and no AR in the remainder. Of 14 patients who had coaptation height assessed in relation to the conduit, 9 patients had type B coaptation and 5 had type C coaptation. (5)

Outcomes

The median stay in the intensive care unit was 7 days. Three patients underwent early aortic valve replacement. One patient with Marfan syndrome and bicuspid aortic valve, who required intraoperative aortic valve resuspension, suffered from severe acute AR 2 days after operation and underwent Bentall surgery. Two patients died in the immediate postoperative period from infectious complications (one case of mediastinitis at 28 days and one case of endocarditis at 47 days). Three patients required a pacemaker implantation because of atrioventricular block. Two patients suffered from endocarditis: one of them had endocarditis by *Klebsiella* that affected the mitral valve, which was complicated by a pseudoaneurysm of the mitral-aortic intervalvular fibrosa 61 days after operation, for which an aortic homograft was placed; the other one had endocarditis by methicillin-resistant *Staphylococcus aureus* that affected the aortic and mitral valves 47 days after operation, for which an aortic homograft was placed.

The median follow-up of 17 patients who underwent Tirone David technique was 4.1 (1.35-12) years, including one patient lost to follow-up. Of the followed-up patients, 43% developed aortic dilatation distal to the implanted conduit. One of them, who had uncharacterized genetic syndrome, underwent replacement of the ascending aorta 4 years after operation and died from dissection of the descending aorta one year after the second surgery.

Coronary button aneurysms were observed in 5 patients.

Two patients had mild-moderate AR, 3 had moderate AR and 2 had severe AR, of whom 1 underwent

Bentall procedure 9 years after VSARR. The rest had mild AR. The higher degrees of regurgitation were caused by leaflet prolapse. The median postoperative Ao annulus size was 21 mm with a Z-score of +2.4 (+1.65 to +2.8). At follow-up, one patient with bicuspid aortic valve disease developed moderate AR.

At present, 81.2% of patients under follow-up are free from reoperation.

DISCUSSION

Aortic root aneurysms are rare in children and adolescents and are mostly associated with connective tissue disorders, (1-4) especially Marfan and Loeys-Dietz syndromes, and bicuspid aortic valve disease. (2,4) Aneurysms have also been described following surgery for conotruncal heart defects. (2) The risk of aortic dissection or rupture is related to the diameter of the aortic root and the ascending aorta. (2-4) Clinical guidelines and surgical indications are based on the progression of dilatation (mm/year), (1) the size of the aortic root and the association with a genetic syndrome.

No guidelines have been established regarding surgical timing in children, so indications are extrapolated from adult recommendations. (5) The risk of these events is very low in this population, and they generally occur in late adolescence. (5,6) In children below the age of 12, the indications for surgery are even less clear, so, in addition to the above, the expression of the genetic syndrome and the degree of AR are considered. (1,4,6) Different groups define criteria for aortic root replacement in children according to their experience (Table 1). In the case of Loeys-Dietz syndrome, Lange et al. consider that Tirone David technique should be postponed until the annulus diameter reaches a value of at least 18 mm so that a prosthetic conduit ≥22 mm may be used and a subsequent reoperation may be avoided. (4)

In adults, the Svensson index is used, which considers the aortic cross-sectional area indexed to height; a value ≥10 implies greater risk of aortic dissection, (7) but this is not validated in children or adults with operated congenital heart disease. (8) Indications for surgery based on aneurysm growth rate >2-3 mm/year may be difficult in children because of the natural growth of the aorta. Davies et al. established the Aortic Size Index (ASI), which is defined as aortic diameter divided by body surface area, to assess the risk of adverse events. An ASI of 2.75-4.25 cm/m² is associated with a moderate risk of rupture (8%/year), whereas an ASI >4.25 cm/m² implies a high risk of rupture (20-25%/year). (6,8,9)

During the last two decades, various surgical techniques have been developed to manage these patients. Aortic root replacement with a mechanical valved conduit (Bentall procedure) was originally proposed, but this procedure involved lifelong anticoagulation, risk of thromboembolism and bleeding. These risks have led to the development of the VSARR procedures, which aim to preserve the aortic valve function and hemodynamics. (4) These procedures include the remodeling or Yacoub technique, and the reimplantation or Tirone David technique. (2) There are many published reports of these techniques in adults, but very few in children. (1) The largest series in this age group which included 100 consecutive cases in 20 years has been reported by Fraser et al. at John Hopkins Hospital. (2) In patients with operated conotruncal heart defects (truncus arteriosus, tetralogy of Fallot, transposition of the great arteries) and in patients who underwent the Ross procedure, who have a very low risk of dissection, aortic root replacement surgery is recommended even in the absence of AR if the diameter is greater than 50 mm and/or the Svensson index is >10 cm/m². (9) In patients with Turner syndrome with an ASI ≥2.5cm/m² and risk factors it is reasonable to perform Tirone David surgery.

Several series have shown that the remodeling technique is not recommended in children because of the high risk of mid-term failure due to the develop-

Tabla 1. Indications for surgery for aortic root replacement

	Marfan	Loeys-Dietz	Bicuspid aortic valve
Fraser et al. (2)	Diam. >5 cm Increase >0.5 cm/year >4.5 cm with family history of rupture >4-5 cm mitral valve surgery	Type I and II: >3.5-4 cm or Z-score >3 Type III: Diam. > 4-4.5cm	Diam. >5.5 cm
Lange et al. (4)	Diam. <5 cm if Z-score >5	Z-score >3	
Kluin et al. (6)	Z-score >4-4.5	Z-score >3 Diam. >3.5cm	
AHA/ACC 2022 Ao Disease Guidelines. (7)	Diam. ≥ 5cm With RF, Diam. ≥ 4.5cm Increase ≥0.3 cm/year Ao root area/height ratio ≥10	Diam. ≥ 4.5cm In at-risk patients, Diam. ≥ 4 cm	Diam. ≥5.5cm Ao root area/height ratio ≥10 With RF, Diam. >5cm

Ao: aortic; Diam: diameter; RF: risk factors

ment of AR following progressive annular dilatation. (2,3,5,6) In contrast, the reimplantation technique is low risk and provides better outcomes, since it offers efficient annular stabilization because of the conduit implantation in the ventriculoarterial junction. The main concern is the stress produced in the leaflets due to the lack of cortex formation, which physiologically occurs in the sinuses. (1) To reduce this stress, conduits with sinuses, such as Valsalva grafts, have been developed, but the smallest size is 24 mm, so they are not appropriate for small children. (4) The long-term performance of the leaflets inside the rigid conduit is still uncertain. Myers et al. have also shown better outcomes in terms of the need for reoperation in patients with operated congenital heart defects using the replacement technique. Besides, they have warned that underestimating the conduit size could result in early valve deterioration. (9)

Regarding the choice of the diameter of the prosthesis, some groups, if possible, postpone surgery until the aortic root size allows the implantation of a conduit for adults. (10) At our institution, we use a conduit diameter that is 3-4 mm larger than the optimal sinotubular junction (STJ) size, as recommended by Cameron and Vricella, to ensure the best apposition of the leaflets. (11) They use conduits that are 2-3 mm larger than the optimal STJ size in patients with Loeys-Dietz syndrome. (12) New techniques for children below the age of 3 have been described using a subannular ring and a superior ring at the STJ to minimize the late onset of AR. (13)

Contraindications for VSARR include marked valve asymmetry, large fenestrations of the leaflets, acute dissection, severe prolapse, valve calcification and ventricularization of the sinuses. (2) According to Baltimore's experience, bicuspid aortic valve is not a contraindication to valve preservation, but special care should be taken to maintain the geometry when resuspending the commissures to ensure postoperative valve competence. (2)

Forty-three percent of patients in our series developed aneurysms in other segments of the aorta distal to the conduit. One patient underwent ascending aorta replacement 4 years after operation, and another underwent arch replacement at the time of the Tirone David surgery. Lange and Fraser suggest reconsidering the management of the distal ascending aorta as well as the aortic arch in patients with severe connective tissue disorders. (2,4) Other groups discourage it because of the high risk of neurological damage (6) and to avoid additional suture lines that may increase the risk of bleeding and pseudoaneurysms.

The iTEE showed trivial residual AR in most patients. During this procedure, we evaluated the presence or absence of AR, the degree of severity, the jet direction and the coaptation height related to the conduit. (14) A patient with Marfan syndrome, bicuspid aortic valve, significant residual AR and eccentric jet underwent reoperation with ECC. Commissural resuspension was performed and his AR remained mild. Two days later, he presented with severe acute AR and underwent Bentall surgery. It has been described that the AR progression could be a consequence of the structural degeneration of the leaflets due to abnormal flow distribution during diastolic closure, or to a functional failure secondary to inadequate coaptation height (less than 11 mm/type C coaptation). (14-17) Five of our patients had moderate or greater AR in the remote postoperative period, and all of them had connective tissue disorders (Table 2). We found no association between the degree of preoperative AR or the presence of bicuspid aortic valve and the development of AR, in contrast with the risk factors described by Tirone David in adult population. (18)

There was no association between the type of postoperative valve coaptation and the development of AR greater than mild at follow-up, in contrast to the study by Hall et al., possibly due to the small sample size. Because of the short follow-up, we are not able to hypothesize whether the cause of AR progression in our patients was due to the degree of leaflet coaptation with respect to the conduit, the impact of the connective tissue disorders, or the valve degeneration. Longer follow-up would be necessary to determine the causes of progression.

In our series, 5 patients developed coronary button aneurysms, as described in the progression of the most severe forms of connective tissue disorders. (9)

At the follow-up of these patients, the recommendation is to include clinical checks and echocardiography every six months during the first postoperative year and then annually, with the addition of tomography

Table 2. Preoperative and postoperative aortic regurgitation.

Degree of AR (n=21)	Preoperative period (n=21)	Immediate postoperative period (n=21)	Remote postoperative period (n=16)
No	2	5	1
Mild	13	16	8
Mild-moderate	1	0	2
Moderate	5	0	2
Severe			3

In the immediate postoperative period, 1 patient died and 3 underwent valve replacement. One patient was lost to follow-up.

or cardiac resonance at least every 2 years. (2,4,10) In the case of Marfan and Loeys-Dietz syndromes, a stricter follow-up is recommended during the first postoperative year with echocardiography every 3 to 6 months. Prescription of beta-blockers and/or angiotensin receptor antagonists should be continued.

The limitations of our study include its retrospective design and the small sample size.

CONCLUSIONS

Children and adolescents with connective tissue disorders or operated congenital heart disease may undergone Tirone David surgery with good outcomes and low mid-term reoperation rate. Neither bicuspid aortic valve nor preoperative aortic regurgitation seems to be associated with greater aortic valve dysfunction at follow-up.

Conflicts of interest

None declared.

(See authors' conflict of interests forms on the web).

Financing

None.

REFERENCES

1. Roubertie F, Ben Ali W, Raisky O, Tamisier D, Sidi D, Vouhé PR, et al. Aortic root replacement in children: a word of caution about valve-sparing procedures. European Journal of Cardio-thoracic Surgery. 2009;35:136-40. https://doi.org/10.1016/j.ejcts.2008.09.043

2. Fraser CD 3rd, Liu RH, Zhou X, Patel ND, Lui C, Pierre AS, et al. Valve-sparing aortic root replacement in children: outcomes from 100 consecutive cases. Ann Thoracic Surgery. 2012;98:299-301.

3. Patel ND, Arnaoutakis GJ, George TJ, Allen JG, Alejo DE, Dietz HC, et al. Valve-sparing aortic root replacement in children: intermediate-term results. Interact Cardiovasc Thorac Surg. 2011;12:415-9. https://doi.org/10.1510/icvts.2010.255596

4. Lange R, Badiu CC, Vogt M, Voss B, Hörer J, Prodan Z, et al. Valve-sparing aortic root replacement in children with aortic root aneurysm: mid-term results. European Journal of Cardio-thoracic. 2013;43:958-64. https://doi.org/10.1093/ejcts/ezs598

5. Vricella LA, Williams JA, Ravekes WJ, Holmes KW, Dietz HC, Gott VL, et al. Early experience with Valve-sparing aortic root replacement in children. Ann Thoracic Surgery. 2005;80:1622-7. https://doi.org/10.1016/j.athoracsur.2005.04.062

6. Kluin J, Koolbergen DR, Sojak V, Hazekamp MG. Valve-sparing root replacement in children. Eur J Cardiothorac Surg. 2016;50:476-81. https://doi.org/10.1093/ejcts/ezw096.

7. 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease. American Heart Association. J Am Coll Cardiol 2022;80:e223-e393.

8. Davies RR, Gallo A, Coady MA, Tellides G, Botta DM, Burke B, et al. Novel measurement of relative aortic size predicts rupture of thoracic aortic aneurysm. Ann Thorac Surg. 2006;81:169-77. https://doi.org/10.1016/j.athoracsur.2005.06.026

9. Buratto E, Konstantinov IE. Valve-sparing aortic root surgery in children and adults with congenital heart disease. J Thorac Cardiovasc Surg. 2021;162:955-62. https://doi.org/10.1016/j.jtcvs.2020.08.116.

10. Rakhra SS, Brizard CP, d'Udekem Y, Konstantinov IE. Valve-sparing aortic root replacement in children. J Thorac Cardiovasc Surg. 2012;144:980-1. https://doi.org/10.1016/j.jtcvs.2012.05.038.

11. Duke C, Luca V. Valve-Sparing Aortic Root Replacement in Children. Operative techniques in thoracic and cardiovascular surgery. 2013;18:2-14. https://doi.org/10.1053/j.optechstcvs.2013.04.001

12. Patel ND, Alejo D, Crawford T, Hibino N, Dietz HC, Cameron DE, et al. Aortic Root Replacement for Children With Loeys-Dietz Syndrome. Ann Thorac Surg. 2017;103:1513-18. https://doi.org/10.1016/j.athoracsur.2017.01.053.

13. Kirklin JK, Johnson WH Jr, Cook BB, Law MA, McMahon WS, Romp RL, et al. Novel technique of valve-sparing aortic root replacement in two children younger than 3 years of age. Ann. Thorac Surg. 2012;94:299-301. https://doi.org/10.1016/j.athoracsur.2012.01.083.

14. Hall T, Shah P, Wahi S. The role of transesophageal echocardiography in aortic valve preserving procedures. Indian Heart J. 2014;66:327-33. https://doi.org/10.1016/j.ihj.2014.05.001.

15. Pethig K, Milz A, Hagl C, Harringer W, Haverich A, et al. Aortic valve reimplantation in ascending aortic aneurysm: risk factors for early valve failure. Ann Thorac Surg. 2002;73:29-33. https://doi.org/10.1016/S0003-4975(01)03312-4

16. David TE, David CM, Feindel CM, Manlhiot C. Reimplantation of the aortic valve at 20 years. J Thorac Cardiovasc Surg. 2017;153:238-9. https://doi.org/10.1016/j.jtcvs.2016.10.081

17. Kachroo P, Kelly MO, Bakir NH, Cooper C, Braverman AC, Kouchoukos NT, et al. Impact of aortic valve effective height following valve-sparing root replacement on postoperative insufficiency and reoperation. J Thorac Cardiovasc Surg. 2022;164:1672-80. https://doi.org/10.1016/j.jtcvs.2022.02.065.

18. David T, Feindel C, David C, Manlhiot C. A quarter of a century of experience with aortic valve-sparing operations. J Thorac Cardiovasc Surg. 2014; 148:872-80. https://doi.org/10.1016/j.jtcvs.2014.04.048