EDITORIAL

The Possibility of Living with a Single Ventricle is Surprising

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My comments on the excellent paper "Total Right Heart Bypass: Long-Term Complications and Survival" by Lafuente et al. (1) performed at the Hospital Garrahan, which is a proud heir of the glorious Hospital de Niños, are accompanied by nostalgic feelings. The article is even more meritorious as it was conducted at a public hospital attended by low-income population (generally living far away from the capital city) which is usually reluctant to answer questionnaires.

The history of total right heart bypass (RHB) started in 1971 when Fontan (2) achieved survival with "ventricularization" of the right atrium (RA) in a tricuspid atresia (TA) by implanting a homograft in the atrial inlet and outlet using a Glenn shunt and closing the atrial septal defect. In the same year, and unaware of Fontan's work, we performed an atriopulmonary anastomosis (APA) in a dying TA (3, 4) leaving a fenestration in the atrial septum and implanting a homograft between the RA and the pulmonary artery. After multiple deliberations with Rodríguez Coronel, Luis Becú and Eduardo Kreutzer, we formulated a different concept from that of Fontan, stating that ventricular end-diastolic pressure (VEDP) is the suction power source necessary to make the system work in the absence of obstruction in the pulmonary circuit. (5)

Later, in 1978, we performed a posterior direct APA (6) behind the aorta, as large as possible, between the right atrium and the main pulmonary artery and its right branch, making a large APA to the appendage roof. This surgical technique became widely used worldwide and corresponds to group I: AP in the article by Lafuente et al. As the authors pointed out, atrial enlargement and arrhythmias can develop between 8 and 12 years after APA. The technique was surpassed in 1988 by the approach proposed by de Leval (7) and Castañeda (8) who developed the lateral tunnel which was not considered in the paper here commented. Then, in 1990 Marceletti (9) described the extracardiac conduit (EC) procedure which is mostly used in all the centers and represents group II: EC in the study by Lafuente et al.

Adult cardiologists should not forget that right atrial pressure, and consequently venous pressure of at least 12 mm Hg is the premise of this peculiar hemodynamic system in which a gradient develops between the RA and the left atrium (LA), whose value depends on VEDP. This is the reason why arrhythmias elevate LA pressure and modify the gradient between both atria, producing clinical problems. The ideal patient would be the one with the lowest values in both atria, with a gradient of approximately 6 mm Hg. In a biventricular patient, high venous pressure values would be interpreted as signs of heart failure. Therefore, life in this hemodynamic system is on the verge of heart failure; however, in the absence of complications, these patients have a near normal life taking low sodium diet for life and under some restrictions. In our experience, we have the longest living patient after total RHB in the world: (10) a 56 year-old woman living a normal life with this system for 40 years.

Firstly, the title of this study seems very adequate, as it reflects the surgery. As the authors state, it is a palliative procedure which the patient or his/her relatives accept as a long-term "mortgage" in return for a reasonable normal life despite having a single ventricle. Who would have dared to affirm before 1971 that it was possible to live with a single ventricle without cyanosis?

The incidence of complications (11, 12) cannot be surprising (58% of total RHB) and is part of the "mortgage" initially acquired. Undoubtedly, this is the best palliative option in patients with single ventricle. On the other hand, "total corrective procedures" for other congenital defects, with the exception of most septal defects, total anomalous pulmonary venous connection and few cases of Jatene surgery, are true palliative procedures with long-term complications despite having two ventricles, requiring exhaustive future surveillance. Of importance, the number of adults already operated on is greater than the number of children waiting for surgery. Therefore, it is necessary to create a public center with specialized cardiovascular surgeons for the management of adults with congenital heart diseases. In our environment, this issue has been partially solved. (13)

The long-term complications of total RHB tend to increase as time goes by depending on the type of single ventricle considered: a powerful left ventricle with

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a normal mitral valve because TA is not the same as other types of single ventricle with abnormal AV valve or other associations.

As the authors have mentioned, the advantage of the EC group over the AP group is the shorter median follow-up (4 years vs. 12 years, respectively). However, the lower incidence of complications particularly arrhythmias in the EC group is reasonable (17% vs. 56%), as the pressure in the single atrium (resulting LA), which depends on VEDP, should be normal and not exerting hypertension on the sinus node and coronary sinus as in the APA. The lower incidence of arrhythmias reported with EC would be due to these factors and to the different X which increases with the years as stated by the authors.

Systemic thrombosis was more common in EC as fenestrations were more frequently performed on the prosthetic material. Other factors as arrhythmias despite anticoagulation, and chronic low cardiac output, (14) could also explain systemic and venous thrombosis.

Protein-losing enteropathy could be related to elevated venous pressure and X.

The development of subaortic stenosis was at least partially solved by the authors with improvement of the surgical technique (Damus-Kay-Stansel procedure or arterial switch).

Diaphragmatic paralysis is a surgical complication that should be carefully prevented. The development of ventricular dysfunction is reasonable if the single ventricle fails. Fortunately, ventricular function improved in 52% of patients after treating the underlying causes.

Interventional cardiologist procedures are of invaluable help to prevent high-risk reoperations. Percutaneous closure of fenestrations is an excellent advantage, especially for EC in which fenestration is more common. Other significant contributions include stent implant to treat venous or pulmonary obstructions, coil occlusion of collateral vessels and closure of patent valves.

Reoperations are due to several causes, including surgical complications as: 1) diaphragmatic paralysis, b) ineffective resolution of a restrictive bulboventricular foramen, c) lack of diagnosis or natural history of valve disease and, d) conversion of APA to EC generally due to badly tolerated arrhythmias. Conversions are more common in adolescents and adults. The number of conversions in this study would be greater if the Garrahan were not a pediatric hospital.

Pacemaker implant is the most common cause of reoperation as this hemodynamic system does not allow ventricular lead placement via the venous system as it happens with biventricular hearts.

The difference in mortality between both groups is not surprising due the difference in X and the technical superiority of EC which reduces the energy loss at the level of the right atrium (14) occurring in APA. In EC, venous velocity is higher because of its lower diameter, as opposed to APA with enlarged RA.

In the discussion, the authors thoroughly address the complications of having a single ventricle previously assumed by the patient or his/her relatives. Undoubtedly, it is essential to preserve ventricular function by treating potential intercurrent conditions. Little can be expected from a dysfunctional single ventricle! and, as the authors pointed out, because ventricular dysfunction was not isolated in any patient, the underlying cause must be treated accordingly. Ventricular function should be preserved with adequate myocardial protection during surgery.

Heart transplantation has not proved to be effective in these patients. (15)

Conflicts of interest

None declared

REFERENCES

1. Lafuente MV, Di Santo M, Mouratian MD, Villalba C, Sciegata A, Saad A y cols. Bypass total del ventrículo pulmonar: complicaciones y sobrevida en el seguimiento alejado. Rev Argent Cardiol 2013;81:XXX-XXX. http://dx.doi.org/10.7775/rac.es.v81.i5.922

2. Fontan F, Baudet P. Surgical repair of tricuspid atresia. Thorax 1971;26:240-8. http://doi.org/d5jjrn

3. Kreutzer G, Galíndez E, Bono H. Una operación para la corrección de la atresia tricuspídea. 5.a Reunión científica de la SAC. Agosto 1971.

4. Kreutzer G, Galíndez E, Bono H, De Palma C, Laura JP. An operation for the correction of tricuspid atresia. J Thorac Cardiovasc Surg 1973;66:613-21.

5. Kreutzer G. Recent Surgical Approach to Tricuspid Atresia. En: Kidd BSL, Rowe RD, editors. The Child with Congenital Heart Disease After Surgery. New York: Futura Publishing Company; 1975. p. 85-97.

6. Kreutzer GO, Vargas FJ, Schlichter AJ, Laura JP, Suarez JC, Coronel AR, et al. Atriopulmonary anastomosis. J Thorac Cardiovasc Surg 1982;83:427-36.

7. de Leval MR, Kilner P, Gewillig M, Bull C, et al. Total cavopulmonary connection: A logical alternative to atriopulmonary connections for complex Fontan operations. Experimental studies and early clinical experience. J Thorac Cardiovasc Surg 1988,96:682-95.

8. Jonas RA, Castaneda AR. Modified Fontan procedure: Atrial baffle and systemic venous to pulmonary artery anastomosis techniques. J Cardiac Surg 1988;3:91-6. http://doi.org/dhprfb

9. Marcelletti C, Corno A, Giannico S, Marino B. Inferior vena cava-pulmonary artery extracardiac conduit: A new form of right heart bypass. J Thorac Cardiovasc Surg 1990;100:228-32.

10. Kreutzer GO. Thirty-two years after total right heart bypass. J Thorac Cardiovasc Surg 2007;134:1351-2. http://doi.org/bptq33

11. Frankin R. The Fontan circulation: Risk factors. En: Redington A, editor. The right heart in congenital heart disease. London: Greenwich Medical Media; 1998. p. 137-44.

12. Kreutzer GO, Schlichter AJ, Kreutzer C. The Fontan/Kreutzer procedure at 40: An operation for the correction of tricuspid atresia. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2010;13:84-90. http://doi.org/d9cv4v

13. Kreutzer C, Klinger D, Cuenca del Rey A y cols. Cinco años de experiencia en el tratamiento quirúrgico de las cardiopatías congénitas del adulto (resumen). Rev Argent Cardiol 2012;80(Supl 3):61.

14. de Leval MR, Dubini G, Migliavacca F, Jalali H, Camporini G, Redington A, et al. Use of computational fluid dynamics in the design of surgical procedures: application to the study of competitive flows in cavopulmonary connections. J Thorac Cardiovasc Surg 1996;111:502-13. http://doi.org/fpz6zq

15. Mitchell MB, Campbell DN, Ivy D, Boucek MM, Sondheimer HM, Pietra B, et al. Evidence of pulmonary vascular disease after heart transplantation for Fontan circulation failure. J Thorac Cardiovasc Surg 2004;128:693-702.