CASE REPORT

Rastelli and Bentall procedures for pulmonary atresia with ventricular septal defect and ascending aorta aneurysm

Jonhy E. Robles-Martínez, MD, and Raymundo García-González, MD.

Departament of Cardiothoracic Surgery, UMAE "Hospital de Cardiología No. 34", IMSS. Monterrey, Nuevo León. MÉXICO.

Pulmonary atresia with ventricular septal defect is a cyanotic congenital heart disease of high complexity, whose association with ascending aortic aneurysm is even rarer. We present the clinical case of this rare association in an adult patient without prior surgical intervention as well as the surgical technique used to correct this pathological condition.

Key words: **Ascending aortic aneurysm; Bentall procedure; Pulmonary atresia; Rastelli procedure; Ventricular septal defect.**

La atresia pulmonar con comunicación interventricular es una cardiopatía congénita de alta complejidad, cuya asociación con aneurisma de aorta ascendente es aún más raro. Presentamos aquí el caso clínico de esta rara asociación en un paciente adulto sin cirugía cardiaca previa, así como los aspectos técnicos empleados para corregir esta patología.

Palabras clave: **Aneurisma de aorta ascendente; Procedimiento de Bentall; Atresia pulmonar; Procedimiento de Rastelli; Comunicación interventricular.**

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Pulmonary atresia with ventricular septal defect (PA + VSD) is defined as the functional absence of continuity between the right ventricle (RV) and the pulmonary arteries, associated with a defect in the ventricular septum that is usually perimembranous. For many authors it constitutes the most severe form of Tetralogy of Fallot. It is, therefore, a cyanogenic heart disease that has the following characteristics: i)decreased development of RV and associated hypertrophy, ii) pulmonary valve atresia with a variable component of hypoplasia and even atresia of the pulmonary arteries, iii) association of a large VSD poorly related by overriding aorta, iv) blood flow, therefore, reaches the pulmonary arteries through the ductus arteriosus in most cases, and on other occasions, through aortopulmonary collaterals, and what makes this heart disease special is how varied the different levels of interruption of the pulmonary pathway can be from the RV to the pulmonary arteries [1].

PA + VSD is a cyanotic congenital heart disease of high complexity, whose association with ascending aortic aneurysm is even rarer. In general, surgical treatment is necessary in childhood, since the general life expectancy without reported surgery is as low as 50% at 1 year, and 8% at 10 years [1].

Corresponding author: Dr. Jonhy E. Robles Martínez
email: jonhy_emanuel@hotmail.com

Ascending aortic dilation and aneurysm formation are not uncommon in conotruncal malformations, such as Tetralogy of Fallot (TOF) and pulmonary atresia with VSD. As the aorta receives most of the blood expelled from both ventricles, the aortic root and ascending aorta can dilate due to hemodynamic stress [2].

Reports of single cases of progressive aortic dilatation long after TOF repair began to appear in the early 1970s. In 1982, Capelli and Somerville mentioned the persistence of a large aortic root, but considered it a characteristic acquired due to long-standing volume overload by years of aortopulmonary shunts before complete repair, and suggested that earlier repair in the first decade of life could prevent this complication [3].

In addition, previous aortopulmonary shunts, congenital anomalies of the aortic valve and acquired infective endocarditis have been reported as cause of aortic regurgitation and, aortic dilation in TOF. Indications for aortic root surgery for these patients differ from Marfan and Loeys-Dietz syndrome due to a low incidence of rupture [2].

Intracardiac anatomy is similar to TOF with pulmonary stenosis. There is a poorly aligned anterior VSD and hypertrophic RV, especially at the level of the infundibulum that generally has no exit. The difficulty of this heart disease is determined by how different the interruption of the pulmo-

nary arteries can be from the RV to the intrapulmonary arteries. When the atrial zone includes the valve and the pulmonary trunk, there may be confluent right and left pulmonary branches joined together. This point is one of the fundamentals approaches to surgical treatment. In patients with confluent pulmonary arteries, a ductus usually persists, through which all lung segments are irrigated [1].

Children with this heart disease usually have clinical manifestations from birth. The most obvious sign is cyanosis. If there are no aortopulmonary collaterals, closure of the ductus will cause severe hypoxemia. Infants, who due to the patent ductus arteriosus (PDA) or the presence of collaterals do not present a florid clinic at birth, progressively present cyanosis and fatigue, impaired growth and cardiac decompensation; all these manifestations, in direct relationship with the greater or lesser pulmonary flow and the growth and activity of the child [1].

The treatment of this heart disease is fundamentally surgical. The ultimate goal of surgery is to close all intracardiac defects, disconnection of all sources of systemic-pulmonary flow, connect the RV and the largest possible number of lung segments: at least 14, but ideally everyone to the central pulmonary artery. The procedures necessary for the treatment of this disease depend on the exact type of anatomy of these patients [1,3].

The natural history of patients with this heart disease is difficult to determine due to the great anatomical variety that these patients present [1].

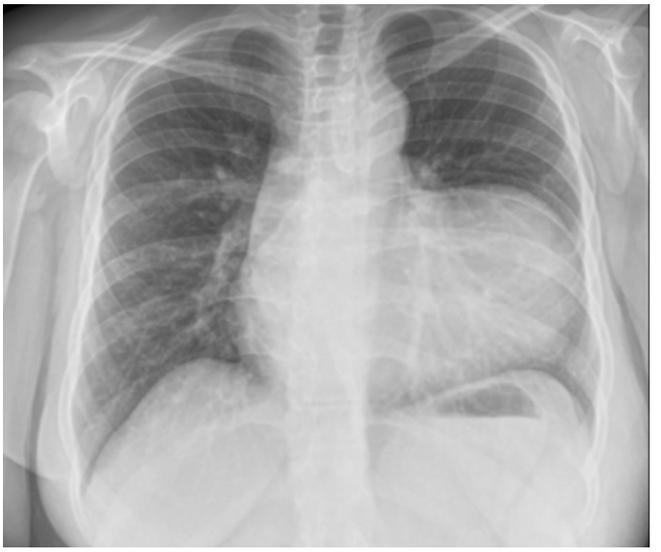

Figure 1. Radiological image in "swedish shoe".

CASE REPORT

A 30-year-old female patient was referred at our institution presenting cyanosis from birth with no previous treatment. Only with periods of dyspnea and mild exacerbations of cyanosis with efforts. She went to medical evaluation at 28 years due to clinical impairment, orthopnea, minimal effort dyspnea, peribucal and distal cyanosis and edema of lower limbs. She just received medical management with beta blocker, diuretics and digoxin.

Study protocol is initiated with a chest radiography observing cardiomegaly and heart in "swedish shoe" (**Fig. 1**). Holter in sinus rhythm leading with left bundle branch block. Transthoracic echocardiogram reported RV hypertrophy, a perimembranous VSD of 18 mm, 50% overriding aorta, and pulmonary valve stenosis with a maximum gradient of 65mmHg, 16mm valve ring, 50% LVEF. TAPSE of 18mm. Angio CT-scan reported aneurysm of ascending aorta with a maximum diameter of 64 mm and in the valvular plane 30 mm. Cardiac catheterization reported dilation of ascending aorta, PASP of 43mmHg, RV pressures of 165/10mm Hg (**Fig. 2**) (**Fig. 3**).

Operation was made through medial sternotomy as usual, on cardiopulmonary bypass. Bicaval approach, myocardial protection with Custodiol anterograde administration. Surgical findings were PA + subaortic VSD of 3 cm in diameter, ASD of 2.5 cm in diameter, 11 mm for PA trunk (Z -4.94), hypoplastic RBPA of 13mm (Z-1.36) with stenosis at the ostium, left branch of pulmonary artery (LBPA) of 11mm (Z -0.85) with confluence stenosis, PDA of 15mm, ascending aortic aneurysm of 7cm in diameter ending just before the first supra-aortic trunk. The operation was performed as follows: PDA ligation was performed with double vascular clip. Then, VSD closure was made with PTFEe patch (**Fig. 4**). Par-

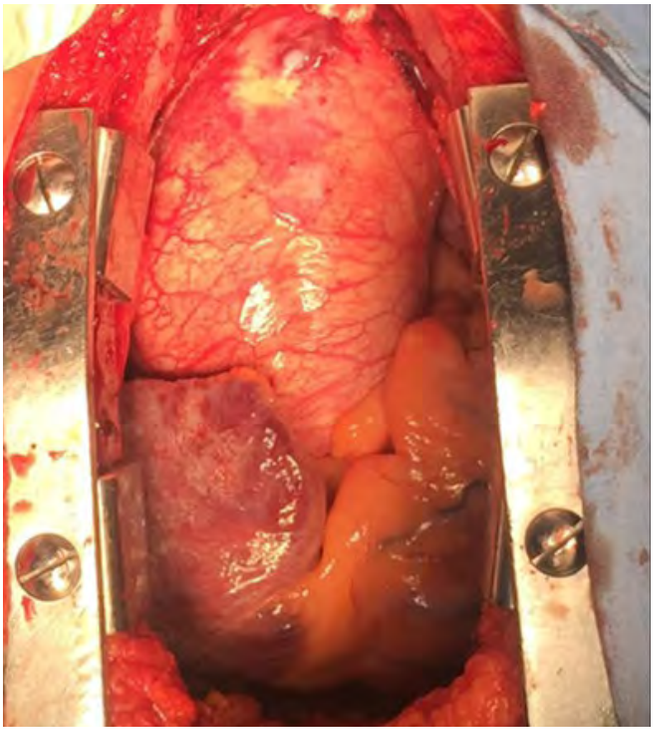

Figure 2. Surgical view of the ascending aorta aneurysm

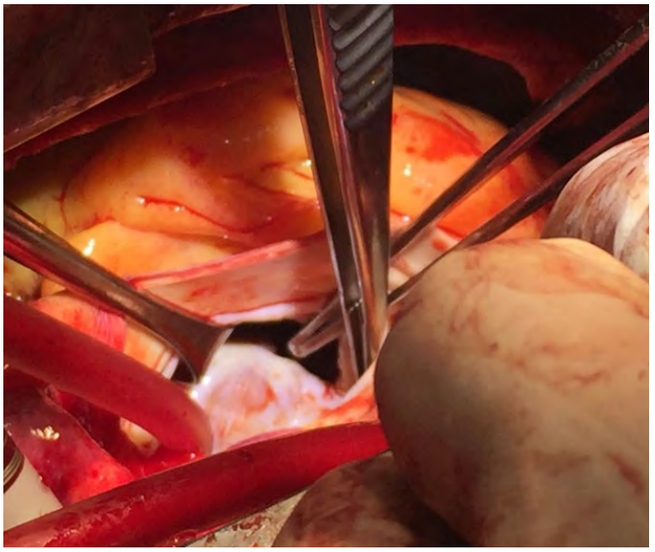

Figure 3. Surgical view: Subaortic ventricular septal defect.

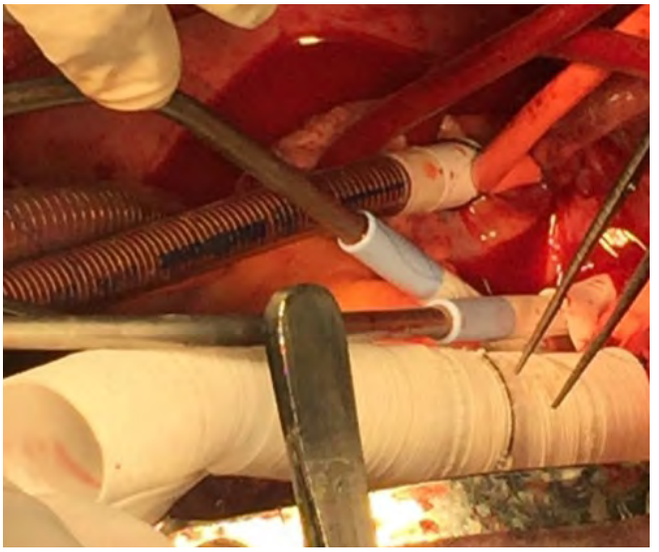

Figure 5. Surgical view: placement of the 22 mm Medtronic Hancock graft

tial ASD closure, resection of aortic aneurysm just above the sinotubular junction. At the same time, enlargement of the right branch of pulmonary artery (RBPA), Rastelli procedure with Medtronic Hancock #22 graft (**Fig. 5**), and ascending aorta replacement with a 30 mm Dacron straight graft (**Fig. 6**) (**Fig. 7**). The extracorporeal circulation time was 318 min, and aortic cross-clamping time was 225 min.

Delay sternal closure was used in this case, being held 48 hours later. Length of stay at ICU was 24 days because of pneumonia associated to mechanical ventilation requiring further tracheostomy. Some bleeding through tracheostomy was observed and bronchoscopies are performed observing bleeding at the right lower lobe bronchus controlled with lidocaine/epinephrine. Control catheterization was performed reporting a 18 mm RBPA, and 12 mm LBPA without aortopulmonary collateral, RV pressure 90/20mmHg. After clinical improvement, she was transferred to the in-hospital general floor. Physical rehabilitation therapy was implemented, in-hospital discharge was after 24 more days under surveillance. Final course has been with no more complications.

COMMENT

At present, complete repair of conotruncal abnormalities is usually preferred with low early mortality during the neonatal or infantile period, and late survival is excellent. How-

Figure 4. Surgical view: closure of the ventricular septal defect with PTFE patch.

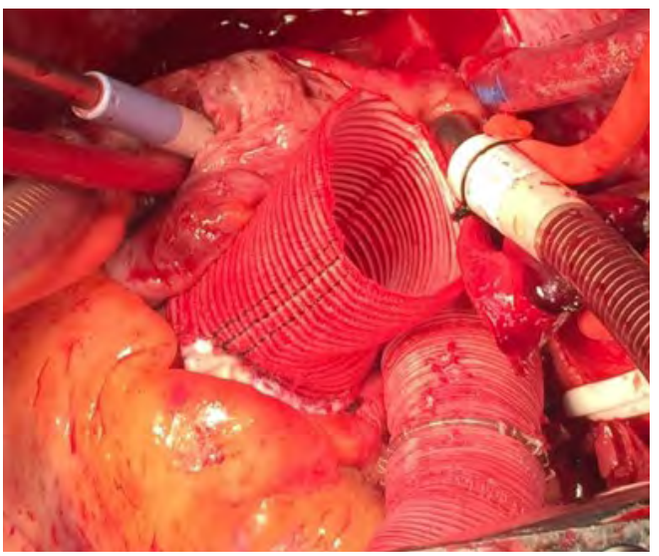

Figure 6. Surgical view: replacement of ascending aorta replacement with a 30 mm Dacron graft.

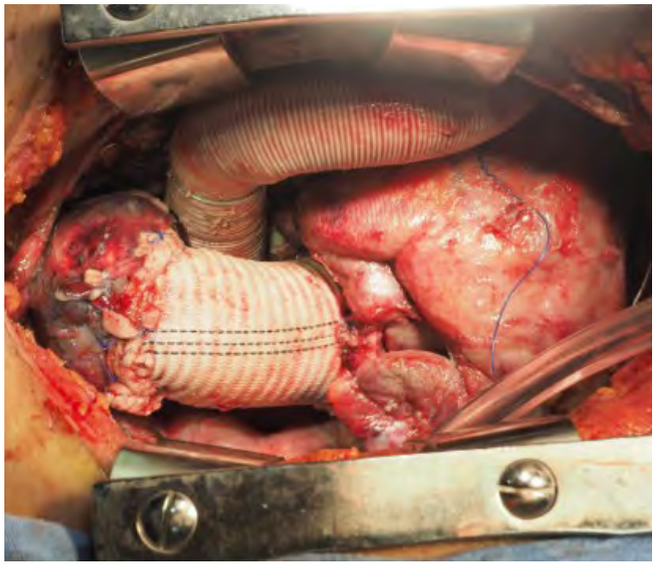

Figure 7. Final aspect of the operatove field, with Dacron graft and Hancock tube in its final position.

ever, many long-term survivors will eventually require at least one reoperation [4]. In the case of our patient, the survival achieved without any surgical treatment during childhood was striking.

Most reports are focused on the reoperative treatment of regurgitant or obstructive lesions on the right-side outflow tract. It is common for the ascending aorta and the aortic root to expand significantly after initial repair of the conotruncal abnormality. Unfortunately, there is little information available on the management of this increasingly frequent problem, and even less information in adult patients who have not received any prior surgical treatment [4].

The pathophysiology of root dilatation in conotruncal abnormalities is probably a combination of increased aortic blood flow before complete repair and vascular intrinsic structural abnormalities [5-7]. It is reasonable to assume that increasing the right-to-left shunt through the VSD with an ascending aortic angulation in the presence of obstruction of the RV outflow tract would lead to aortic dilation due to volume overload [6]. However, the question about whether the progressive enlargement of the ascending aorta is due to an inherent or acquired aortopathy remains unanswered. This is due to what was observed in patients operated after 7 years in whom aortic root dilation progressed.

The aortic arch of the right side, the male gender, history of aortopulmonary shunt, and complete repair at an older age have been associated with late aortic dilation [3].

There is just a few evidence in the literature about the indications for surgical intervention in the aortic root in conotruncal anomalies. Current consensus recommendations for adult patients with congenital heart disease are to repair the ascending aorta when it is larger than 55 mm in diameter, as in the case of our patient. Others have suggested intervention if there is moderate or severe aortic regurgitation [4].

In conclusion, we can say that there are just a few reports of patients treated surgically in adulthood. The anatomical complexity of this heart disease requires the exact knowledge of the disease and congenital defects of each patient. Surgical management is complex and must be individualized. Meanwhile, we should wait for more further information coming from expert groups and specialized centers in order to get better outcomes and conclusions as well.

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REFERENCES

1. Centella-Hernández T, Stanescu D y Stanescu S. Atresia pulmonar con comunicación interventricular. Cir Cardiov 2014;21:132-7.
2. Chiostri B, Piccinini F, Vrancic M, et al. Combined Rastelli and Bentall procedures for pulmonary atresia with ventricular septal defect and a large aortopulmonary collateral artery and ascending aorta aneurysm. World J Pediatr Congenit Heart Surg 2014;5:467-9.
3. Francois K. Aortopathy associated with congenital heart disease: A current literature review. Ann Pediatr Cardiol 2015; 8: 25-36.
4. Dearani JA, Burkhart HM, Stulak JM, Sundt TM, Schaff HV. Management of the aortic root in adults patients with conotruncal anomalies. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009; 12:122-9.
5. Karamlou T, McCrindle BW, Williams WG. Surgery insight: Late complications following repair of tetralogy of Fallot and related surgical strategies for management. Nat Clin Pract Cardiovasc Med 2006; 3:611-22.
6. Niwa K, Siu SC, Webb GD, Gatzoulis MA. Progressive aortic root dilatation in adults late after repair of tetralogy of Fallot. Circulation 2009;106:1374-8.
7. Tan JL, Gatzoulis MA, Ho SY. Aortic root disease in tetralogy of Fallot. Curr Opin Cardiol 2006;21:569-72.