Aortopulmonary Window: Clinical Assessment and Surgical Results

Antonio Moruno Tirado, José Santos de Soto, Josefina Grueso Montero, José L. Gavilán Camacho, Antonio Álvarez Madrid[a], Mauro Gil Fournier[a] and Alfonso Descalzo Señorans

Secciones de Cardiología y Hemodinámica. [a]Servicio de Cirugía Cardiovascular. Hospital Infantil Virgen del Rocío. Sevilla.

Introduction and objectives. Aortopulmonary septal defect is an uncommon congenital cardiac anomaly. To date, approximately 300 cases have been reported. We present our experience, emphasizing the importance of early correction to avoid irreversible pulmonary hypertension.

Patients and method. Between 1979 and 2000, seven patients underwent surgical repair of this heart defect in our hospital. Two had type I (proximal), 4 had type II (distal) and 1 had type III (complete). Complex associated cardiac anomalies were present in 4 cases: type A interruption of the aortic arch in 2 cases, hypoplastic aortic arch in 1 and transposition of great arteries with ventricular septal defect in 1.

Four cases (57%) were diagnosed by echocardiography. In all patients diagnoses were confirmed by cardiac catheterization.

Patient records were reviewed retrospectively, with special attention to clinical, echocardiographic and hemodynamic data as well as surgical characteristics.

Results. No intraoperative deaths occurred. The patient with associated transposition of great arteries died 22 days after surgery as a result of severe pulmonary hypertension. The remaining patients are asymptomatic without treatment after a mean follow-up period of 69 months.

Conclusions. Even though aortopulmonary septal defect is a rare anomaly, it should be considered wheneverthe course of complex congenital heart disease includes early cardiac failure and pulmonary hypertension. Repair before 6 months will prevent irreversible damage of pulmonary vessels.

Key words: *Aortopulmonary window. Diagnosis. Early correction.*

Ventana aortopulmonar: valoración clínica y resultados quirúrgicos

Introducción y objetivos. El defecto del septo aortopulmonar es una cardiopatía congénita rara. Hasta el momento se han publicado alrededor de 300 casos. Presentamos nuestra casuística, resaltando la importancia de su corrección precoz para evitar el desarrollo de enfermedad vascular pulmonar irreversible.

Pacientes y método. Entre 1979 y 2000, 7 pacientes fueron intervenidos de dicha cardiopatía en nuestro centro. Dos pacientes presentaron el tipo I (proximal), cuatro el tipo II (distal) y uno el tipo III (defecto completo del septo aortopulmonar). En 4 casos (57%) se asociaron anomalías complejas: interrupción del istmo aórtico tipo A (n = 2), hipoplasia del istmo aórtico (n = 1) y transposición de los grandes vasos con comunicación intraventricular.

El diagnóstico se realizó mediante ecocardiografía en 4 casos (57%) y en todos fue confirmado mediante estudio hemodinámico.

Realizamos un estudio retrospectivo de sus historias clínicas, con especial atención a los datos clínicos, del diagnóstico ecocardiográfico y hemodinámico, así como a los aspectos quirúrgicos.

Resultados. No hubo ningún fallecimiento durante la intervención quirúrgica. El enfermo portador de transposición de los grandes vasos con comunicación interventricular falleció 22 días después a causa de hipertensión pulmonar severa. Tras un seguimiento medio de 69 meses, los enfermos restantes se encuentran asintomáticos sin medicación.

Conclusiones. A pesar de su rareza, se debe pensar en esta entidad ante toda cardiopatía compleja que curse con insuficiencia cardíaca e hipertensión pulmonar precoces. La corrección antes de los 6 meses evitará el desarrollo de enfermedad vascular pulmonar irreversible.

Palabras clave: *Ventana aortopulmonar. Diagnóstico. Cirugía.*

INTRODUCTION

The aortopulmonary window (APW) is a communication between the ascending aorta and the pulmonary trunk and/or right pulmonary branch, in the presence of both separate sigmoid valve planes, which

---

ABBREVIATIONS

ECC: extracorporeal circulation
AoAI A: type A aortic arch interruption
TA: truncus arteriosus
APW: aortopulmonary window

---

differentiates it from truncus arteriosus (TA).

This rare anomaly represents 0.2%-0.6% of all congenital heart diseases.[1] Since it was first described in 1830[2] until the present, only about 300 cases have been published, for the most part as isolated reports.[3-5] Half of the patients have other more or less complex associated cardiac defects (complex windows) that make the diagnosis difficult.

Different modalities of APW exist and several classifications have been proposed. The classification most often used is that of Mori et al,[6] which divides them in type I, or proximal (70%) (the defect is circular, located in a zone equidistant between the sigmoid valve plane and the pulmonary bifurcation); type II, or distal (25%) (of spiral form, it affects the trunk and origin of the right pulmonary artery), and

type III (5%) (complete defect of the aortopulmonary septum).

In general, these malformations originate an important left-right shunt with congestive heart failure in the first days or months of life, and early development of severe pulmonary hypertension.[7]

We report our experience with 7 patients diagnosed and treated surgically in our center.

PATIENTS AND METHOD

Between 1979 and 2000, 7 patients underwent surgery for APW. Four were females and 3 were males, the mean age at the time of the intervention was 8 months (range, 11 days to 30 months), and the mean weight was 6.2 kg (range, 2.7-14.8 kg).

All patients underwent a clinical study, chest radiograph, electrocardiogram, cardiac echocardiography, and catheterization (Table 1).

The first two patients were asymptomatic and had been referred to the hospital to study a systolic murmur of moderate intensity located in the upper left parasternal area. In the clinical examination, electrocardiogram, and chest radiograph, the findings were suggestive of left-right shunt. The third patient suffered heart failure at the 3 months of life precipitated by a pulmonary infection. The other

TABLE 1. **Echocardiographic and hemodynamic clinical data**

Demographic data				Clinical presentation	Echocardiography	Hemodynamic study				
Case	Age	Sex	Weight			Window type	Associated anomaly	AP pressure	Qp/Qs	RVP
1/1984	30 months	M	14.5	Study of murmur L-R shunt symptoms	Ductus Dilation of left cavities	L (simple)		25/10	2.1 (52%)	1.32
2/1995	17 months	M	10	Study of murmur L-R shunt symptoms	Ao-P window	II (simple)		80/38	1.7 (41%)	3.81
3/1998	3 months	V	5.4	Heart failure Pulmonary infection Continuous murmur	Large ductus Pulmonary hypertension	III (simple)	55/34	7.25(86%)	1.5	
4/1993	7 days	M	2.5	Heart failure Continuous murmur	D-TGV VSD Ao-P window Pulmonary hypertension	I (complex)	D-TGV VSD	65/30	4.5 (84%)	3.4
5/1991	7 days	V	3.4	Heart failure No femoral pulses Murmur on back	Ao-P window Critical Co-Ao Pulmonary hypertension	II (complex)	Type A AoAI	80/44	4.1 (75%)	4.6
6/1992	10 days	M	3.5	Cardiogenic shock No femoral pulses	Ao-P window AoAI Pulmonary hypertension	II (complex)	Type A AoAI	85/24		
7/2000	2 days	V	3.4	Heart failure No femoral pulses Murmur on the back	Hypoplastic isthmus Ao RSA OS Pulmonary hypertension	II (complex)	AoI hypoplasia SA OS Raberrant RSA	60/24	3.73 (73%)	4.5

Ao-P indicates aortopulmonary; RSA, right subclavian artery; Co-Ao, coarctation of the aorta; AoAI, aortic arch interruption; AoI, aortic isthmus; L-R, left-right; OS, ostium secundum.

patients, who had complex windows, showed symptoms of severe heart failure in the first 10 days of life (case 6, cardiogenic shock).

In only 2 cases (3 and 4) was a continuous precordial murmur auscultated, which is described as somewhat lower than that corresponding to ductus and characteristic of the disease.

In 6 patients extracorporeal circulation (ECC) was used and the APW was approached by aortotomy and closed with an internal PTFE patch. In complex windows, the associated anomaly was corrected in the same surgical act. In case 4, closure was by direct suture without EEC, at the same time that the pulmonary artery was banded.

We evaluated the clinical aspects, echocardiographic diagnosis, and their correlation with the hemodynamic study that all patients underwent, as well as the incidents that occurred in the immediate postoperative period and later follow-up.

RESULTS

In Table 1, the clinical, echocardiographic and hemodynamic data of the patients are collected.

The diagnosis was initially made by echocardiography in 4 patients (57%). In case 7, a second echocardiography was made, which was indicated when the patient did not improve after a previous correction of severe coarctation of the aortic isthmus.

In all patients, the diagnosis was confirmed by hemodynamic study. Except for case 1, the patients presented severe pulmonary hypertension with pulmonary pressures that were practically the same as systemic pressures. The left-right shunt was voluminous in complex windows, reaching 86% of pulmonary flow in the absence of the entire aortopulmonary septum (case 3).

Two patients had type I APW, four type II, and one type III.

Four patients (57%) also presented an associated complex cardiac anomaly: type A (AoAI A) interruption of the aortic arch in 2 cases, hypoplasia of the aortic isthmus in one, and D-TGV with VSD in the one. These patients underwent surgery in the first month of life, when they had a mean weight of 3.6 kg (range, 2.7-3.9 kg).

No patient died during the intervention. Patient 4 died 22 days after surgery due to pulmonary hypertension refractory to treatment. Patients 3 and 6 presented minor complications in the immediate postoperative period.

After a mean period of follow-up of 6 years (range, 1 month to 16 years), the patients are asymptomatic without requiring medication and lead normal lives, except for patient 5, who has a moderate maturational delay. During the intervention, this patient had the longest aortic clamping time, reaching 75 min.
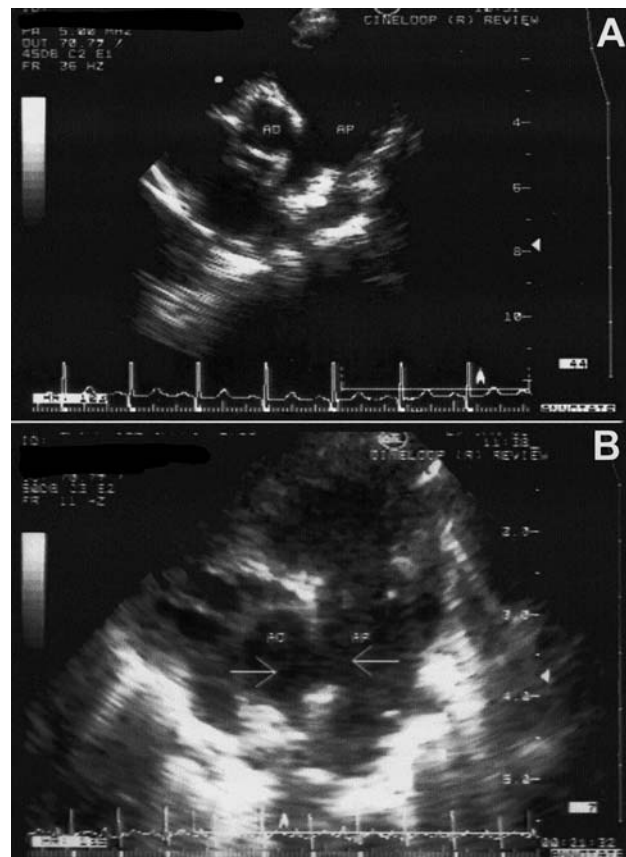

Fig. 1. Bidimensional echocardiography in the cross-sectional parasternal plane of the great vessels corresponding to 2 different patients: A, true type II APW. B, false APW. Although the defect seems clear, including the presence of the T sign, the hemodynamic study revealed a large ductus instead of APW. On this echocardiographic plane, the diagnosis of APW must be made cautiously.

DISCUSSION

APW can result from incomplete fusion (type I), poor alignment (type II), or total absence (type III) of the right and left conotruncal cushions, which normally complete conotruncal septation between weeks 5 and 8 of intrauterine life.[8] Recent studies indicate that it does not represent an earlier stage of TA, as had been generally accepted, but that the two anomalies have different origins. The origin of the latter malformation lies in a disturbance of the cells from the neural crest.[1]

In contrast with reports in the literature,[6,9] in our series the incidence of type II windows was greater (57%) than that of type I (28%). We had one type III case, which is exceptional.

In half of the patients, APW is associated with other cardiac defects, the most frequent being: AoAI in 15-20% (principally type A), ductus arteriosus in 11%, VSD in 8%, coronary anomalies in 8%, tetralogy of Fallot in 5%, ect.[10] AoAI or severe
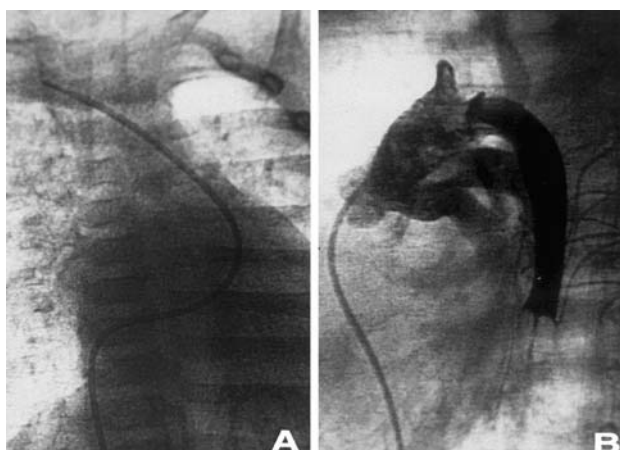
Fig. 2. Hemodynamic APW study. A: Right catheterization. Through the APW the catheter was advanced from the pulmonary artery to the ascending aorta and brachiocephalic trunk. B: Stop flow aortography, lateral view. The two vessels filled simultaneously in the presence of 2 separate sigmoid valves without the existence of ductus arteriosus.

preductal coarctation of the aorta (considered different stages of same organization[11]) is very rare as an isolated disease. The high incidence found in association with APW could be attributed to the notable decrease in blood flow that the aortic isthmus suffers during the prenatal period in these cases. In TA, the association with type B AoAI is much more common, which coincides with an etiopathogenesis different from that of APW, as mentioned previously.

Until the moment, to our knowledge, only 3 cases of APW associated with TGV have been reported:[12,13] the first two died, as did our patient, which is the fourth case communicated in the literature.

Both the type of associated lesion and size of the APW condition the clinical manifestations of patients. The diagnosis must be suspected in cases of early heart failure with signs of significant left-right shunt, such as dilation of the left cavities, particularly the left atrium, and/or functional mitral insufficiency with a morphologically normal valve,[14] associated with severe early pulmonary hypertension. The continuous cardiac murmur characteristic of the disease is auscultated in fewer than half of the cases, as occurred in our cases.

The electrocardiographic and radiological findings are non-specific, which is why echocardiography has an important role in diagnosis. The cross-sectional parasternal planes over the aortic valve plane (Figure 1), coronal subcostal plane of both outflow tracts, suprasternal longitudinal and upper parasternal planes are used.

Some false positives are found, particularly when using equipment with scant lateral resolution, because there may be an artificial echo-loss phenomenon (dropout) in the region of the aortopulmonary septum,

due to the alignment of the septum in the direction of the lateral resolution of the transductor.[15] To differentiate this phenomenon from true APW, aside from exploring the septum in several planes, some authors resort to the T sign. This sign is nothing other than the greater refringence that the edge of the true defect acquires, which is perpendicular to the rest of the septum and adopts a T image.

The Doppler color study is of inestimable aid and reveals a low-speed bidirectional laminar flow in large, unrestricted defects, with pulmonary hypertension and continuous turbulent flow in the trunk and/or right pulmonary artery. A high-speed flow without pulmonary hypertension is found in small defects.

In our experience, neither the T sign nor Doppler color images provide a definitive diagnosis, as shown in Figure 1. In our series, the diagnostic performance of echocardiography was 57% (4/7), in consonance with reports: 53% (70% in simple APW and 37% in complex APW).[16] In patient 1 (1984) we did not have Doppler color, and in patient 7 an echocardiographic diagnosis was made in the second study, which was performed when the patient did not progress well after a correction was made without a previous hemodynamic study of the associated defect. As reported in the literature, false diagnoses, both positive and negative, are not infrequent. Negative diagnoses are related mainly to the existence of associated complex anomalies that explain the symptoms of the patient. For this reason, we believe that the diagnosis should be confirmed by cardiac catheterization in all cases (Figure 2).

Due to the rapid development of irreversible pulmonary vascular disease, the malformation should be repaired when diagnosed, preferably before 6 months of age. When total pulmonary vascular resistance at the time of the intervention is <8 U/m^2, the long-term evolution is optimal, as occurred in all our surviving patients.

Since the first correction made by Gross,[17] numerous techniques have been described, with or without ECC, and using a transaortic or transpulmonary patch. The transaortic approach is preferred because it provides better exposure of the window and ostium of the left coronary artery.[18] Matsuki et al[19] and Meissner[20] later described the use of a pulmonary artery flap, later using autologous pericardium to repair the pulmonary artery. Di Bella et al[21] also published a similar technique, but without using a patch to repair the pulmonary artery, which was closed using aortic adventitia, which produces excellent results when autologous tissues with normal growth potential are used.

Some cases of percutaneous closure with favorable results have been reported in specific situations, such as type I, small (3-4 mm) windows not associated with other anomalies, particularly in the origin of the

coronary arteries.[22,23]

CONCLUSIONS

Echocardiography should be the diagnostic method of choice, but it does result in some false positive and negative diagnoses. For this reason, we still confirm the diagnosis by hemodynamic study.

Both the diagnosis and the correction must be made before the age of 6 months to avoid the development of irreversible pulmonary vascular disease.

DEDICATION

To Dr. Salud Ariza Almeida, Head of the Cardiology Section, Hospital Infantil Virgen del Rocío of Sevilla. *In memoriam.* With affection from all of us who were once her residents.

REFERENCES

1. Kutche LM, Van Mierop LHS. Anatomy and pathogenesis of aortopulmonary septal defect. Am J Cardiol 1987;59:443-7.
2. Elliotson J. Case of malformation of the pulmonary artery and aorta. Lancet 1830;1:247-51.
3. Tkebuchava T, von Segesser LK, Vogt PR, Bauersfeld U, Jenni R, Kunzli A, et al. Congenital aortopulmonary window: diagnosis, surgical technique and long-term results. Eur J Cardiothorac Surg 1997;11:293-7.
4. McElhinney DB, Reddy VM, Tworetzky W, Silvermann NH, Hanley FL. Early and late results after repair of aortopulmonary septal defect and associated anomalies in Infants < 6 month of age. Am J Cardiol 1998;81:195-201.
5. Brook MM, Heymann MA. Aortopulmonary window. En: Allen HD, Clark EB, Gutgesell HP, Driscoll DJ, editors. Moss and Adams' heart disease in infants, children and adolescents. Philadelphia: Lippincott Williams and Wilkins, 2001;p.670-4.
6. Mori K, Ando M, Takao A, Ishikawa S, Imai Y. Distal type of aortopulmonary window. Br Heart J 1978;40:681-9.
7. Van Son JA, Puga FJ, Danielson GK, Seward JB, Mair DD, Schaff HV, et al. Aortopulmonary window: factors associated with early and late success after surgical treatment. Mayo Clin Proc 1993;68:128-33.
8. Soares AM, Atik E, Martins Cortez T, Albuquerque AM, Castro CP, Barbero-Marcial M. Janela aortopulmonar: análise clínico-cirúrgica de 18 casos. Arq Bras Cardiol 1999;73:59-66.
9. Tiraboshi R, Salomone G, Cruppi G, Manasse E, Salim A, Carminati M, et al. Aortopulmonary window in the the first year of life: report on 11 surgical cases. Ann Thorac Surg 1988;46:438-41.
10. Faulkner LS, Olhan RR, Atwood GF, Graham TP Jr. Aortopulmonary window, ventricular septal defect and membranous pulmonary atresia with a diagnosis of truncus arteriosus. Chest 1974;65:351-3.
11. Van Mierop LHS, Kutsche LM. Interruption of aortic arch and coarctation of the aorta: pathogenetic relations. Am J Cardiol 1984;54:829-34.
12. Krishnan P, Airan B, Sambamurty MS, Shrivastava S, Rajani M, Rao IM. Complete transposition of great arteries with aortopulmonary window: surgical treatment and embriologic significance. J Thorac Cardiovasc Surg 1991;101:749-51.
13. Marangi D, Peterson RJ, Ceithaml EL, Marvin WJ Jr. Surgical repair of D- transposition with aortopulmonary window: a case report. J Thorac Cardiovasc Surg 1996;111:671-2.
14. Comin J, Manito N, Roca J, Castells E, Esplugas E. Insuficiencia mitral funcional. Fisiopatología e impacto del tratamiento médico y de las técnicas quirúrgicas de reducción ventricular izquierda. Rev Esp Cardiol 1999;52:512-20.
15. Snider AR, Serwer GA, Ritter SB. Defects in cardiac septation. En: De Young L, Geistler JB, Patterson J, editors. Echocardiography in pediatric heart disease. St Louis: Mosby-Year Book Inc., 1997; p.292-3.
16. Balaji S, Burch M, Sullivan ID. Accuracy of cross-sectional echocardiography in diagnosis of aortopulmonary window. Am J Cardiol 1991;15:650-3.
17. Gross RE. Surgical closure of an aortic septal defect. Circulation 1952;5:858-63.
18. Castañeda AR, Jonas RA, Mayer JE, Hamley FL. Cardiac surgery of the neonate and infant. Philadelphia: WB Saunders 1994; p.295-300.
19. Matsuki O, Toshikatsu Y, Yamamoto F, Nishigaki K, Uemura H, Kawashima Y. New surgical technique for total defect aortopulmonary window. Ann Thorac Surg 1992;54:991-2.
20. Messmer BJ. Pulmonary artery flap for closure of aortopulmonary window. Ann Thorac Surg 1994;57:498-501.
21. Di Bella I, Gladstone DJ. Surgical management of aortopulmonary window. Ann Thorac Surg 1998;65:768-70.
22. Stamato T, Benson LN, Swalhorn JF, Freedom RM. Transcatheter closure of an aortopulmonary window with a modified double umbrella occluder system. Cath Cardiovasc Diagn 1995;35:165-7.
23. Jureidim SB, Spadioro JJ, Rao OS. Successful transcatheter closure with the buttored device of aortopulmonary window in an adult. Am J Cardiol 1998;81:371-2.