CASE REPORT
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Aortopulmonary window: diagnosis in the operating room

Ventana aortopulmonar: diagnóstico en quirófano

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ABSTRACT

Aortopulmonary window is a rare congenital defect that affects about 0.2% of the population. It is diagnosed and corrected during the first weeks of life. In adulthood it is rare and involves a high mortality. We present the case of a 17-year-old female, since the first years of life presents distal cyanosis and systolic murmur. Fixed fibrous subaortic stenosis, ventricular septal defect and severe mitral insufficiency were diagnosed. Surgical resolution was performed where aortopulmonary window was diagnosed in the operating room.

Keywords: congenital defect, aortopulmonary window, pulmonary hypertension.

RESUMEN

La ventana aortopulmonar es un defecto congénito infrecuente que afecta alrededor de 0.2% de la población. Se diagnostica y corrige durante las primeras semanas de vida. En la edad adulta es rara e implica una alta mortalidad. Presentamos el caso de paciente femenino de 17 años, desde los primeros años de vida presentó cianosis distal y soplo sistólico. Se diagnosticó estenosis subaórtica fibrosa fija, comunicación interventricular e insuficiencia mitral severa. Se realizó resolución quirúrgica donde se diagnosticó ventana aortopulmonar en el quirófano.

Palabras clave: *defecto congénito, ventana aortopulmonar, hipertensión pulmonar.*

Abbreviations:
APW = aortopulmonary window
CT = computed tomography
ECOTT = transthoracic two-dimensional echocardiography
FSSSA = fixed fibrous spur type subaortic stenosis
PDA = persistent ductus arteriosus

INTRODUCTION

Aortopulmonary window (APW), also referred as aortopulmonary septal defect, is a rare congenital heart defect. It is defined as abnormal communication between ascending aorta and pulmonary trunk, with the presence of separate semilunar valves.[1] Regarding the incidence of this defect, several authors present different figures ranging from 0.1-0.25%.[2] Others suggest figures of 0.2 to 0.6% of all congenital heart defects.[3]

Presentation of APW during adulthood is usually rare, being surgically corrected during early childhood.[1] If surgical repair is not performed during the first months of life, mortality in the first year of life is between 40 and 50%.[4] Patients in whom incidental diagnosis of APW is found during adulthood often present with pulmonary hypertension.[5] The progression of the disease is caused by increased pulmonary flow due to the presence of left-right shunt, generating irreversible pulmonary vascular obstructive disease, as well as congestive heart failure symptoms and the development of Eisenmenger's syndrome.[6]

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The earliest classification of APW was described in 1978 by Katsuhiko Mori in a series of 14 cases. This paper provided a classification of three types.[7] Subsequently, in 2000, members of the STS-Congenital Heart Surgery Database Committee and representatives of the European Association for Cardiothoracic Surgery took up the classification established by Mori in 1978 to establish a new nomenclature for APW. Currently, 4 types of aortopulmonary septal defects have been described: type 1 or proximal defect. In the posteromedial wall of the ascending aorta, over the sinus of Valsalva, a few millimeters above the semilunar leaflets. With a sufficient superior border, but a few millimeters inferior border, which separates the APW from the semilunar valves. Type 2 or distal defect. This type of APW location is situated in the superior portion of the ascending aorta. It presents a sufficiently large inferior border and a scarce superior border. Type 3 or total defect. It is a defect that extends over most of the ascending aorta, with poorly marked superior and inferior borders. Type 4 or intermediate type. It is a defect that extends mostly in the portion of the ascending aorta, like type 3 or total defect, but presents superior and inferior borders of adequate size.[8]

The aortopulmonary septal defect can be isolated, meaning that APW alone is present, or complex and accompanied by other cardiac defects. It has been reported that the association with other defects is 25 to 35%.[9] Some of them are persistent ductus arteriosus (PDA), interrupted aortic arch, severe coarctation, tetralogy of Fallot, aortic atresia, and ventricular septal defects.[10] Coexistence of interrupted aortic arch is more frequently found.[11]

The diagnosis is made by transthoracic ultrasound with Doppler, which is key and confirmatory for the detection of the defect.[2] This defect may go undetected by ultrasound, so other imaging tests may be used to confirm the diagnosis such as computed tomography angiography (CT) or cardiac catheterization. Once the diagnosis is established, it is important that correction of the defect is performed surgically as early as possible. Several studies have shown that closure using a patch through a transaortic approach is possible.[12]

Subaortic stenosis, also called subvalvular aortic stenosis, is a congenital heart disease resulting from the formation of fibromembranous tissue.[13] It is one of the most common cardiopathies in the aorta, with a prevalence of 6.5%; it is the second most common type of aortic stenosis. This formation of fibrotic tissue generates an increased pressure gradient in the outflow tract of the left ventricle due to a fixed obstruction in blood flow. It is associated with other defects such as ventricular septal defect, patent ductus arteriosus, coarctation of the aorta, bicuspid aortic valve, abnormal left ventricular papillary muscle and/or atrioventricular septal defect, among others.[14] The diagnosis is made by echocardiogram. Clinically they remain asymptomatic; however, symptoms can appear when performing activities that generate physical stress or during pregnancy.[10]

CASE DESCRIPTION

We present the case of a 17-year-old female patient, whose pathology began at birth, with development of cardiac symptoms, presenting distal cyanosis and heart murmur without follow-up and without treatment. She began her follow-up until the age of 13 years when she went to the health center for presenting grade III malnutrition, where

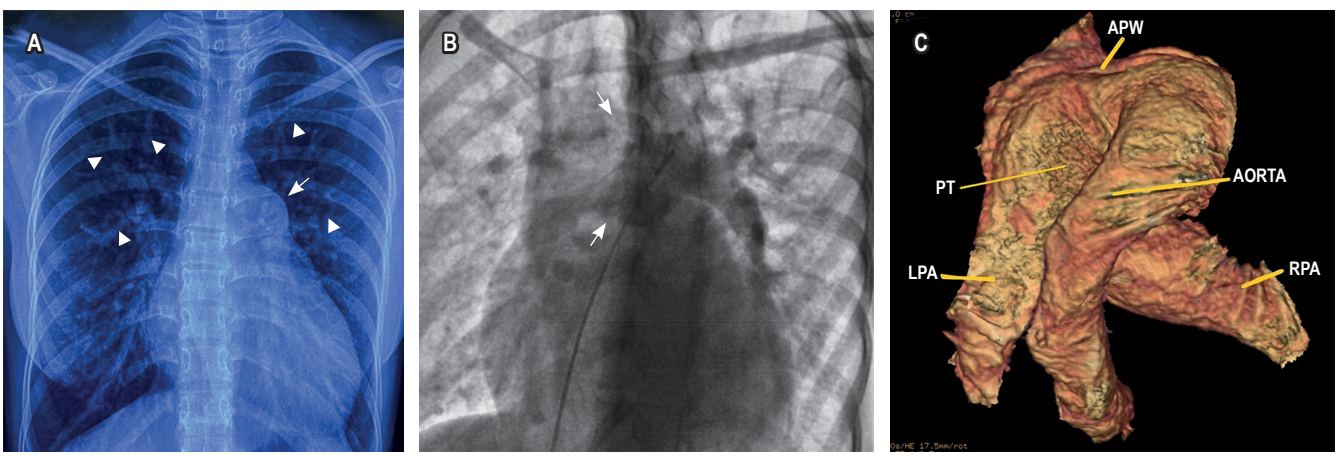

Figure 1: A) Chest X-Ray performed prior to surgery, showing dilatation of the pulmonary artery trunk (arrow), increased pulmonary flow, which identifies the increase of the bronchovascular network by veno-capillary congestion in both hemithorax (arrowhead). **B)** Pre-surgical diagnostic right catheterization where the filling phase of the pulmonary artery is identified where a slight image of leakage through the APW is identified. **C)** Angiotomography, pre-surgical reconstruction of great vessels where communication between the pulmonary and aorta is identified.
APW = aortopulmonary window. LPA = left pulmonary artery. PT = pulmonary trunk. RPA = right pulmonary artery.

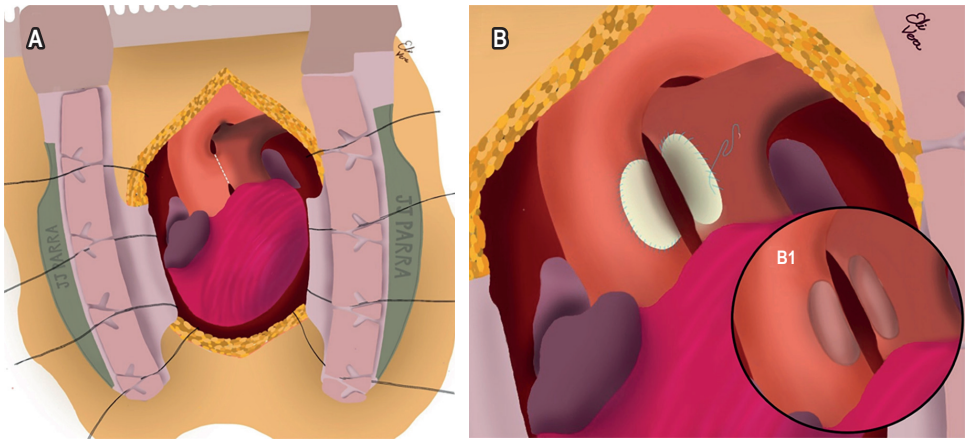

Figure 2:

A) Representation of median approach before median sternotomy. Incision is made over the anterior wall of the defect. **B)** Illustration depicting the separation of the structures and placement of pericardial patch on each structure.

she was evaluated and sent to pediatric cardiology due to the presence of a murmur.

At 14 years of age, she was evaluated annually by pediatric cardiology with suspicion of congenital heart disease. On physical examination, she presented as acyanotic with no evidence of respiratory distress. Auscultation at the precordial level identified a grade II/VI systolic ejection murmur in the second intercostal space, increased volume in the left hemithorax, normal pulses in the extremities. Cardiomegaly grade I at chest X-Ray. Electrocardiogram with no important alterations.

During follow-up, at the age of 17 years there was clinical progression with the development of dyspnea on moderate efforts. Auscultation identified progression of systolic murmur to grade III/VI. A chest X-Ray was performed showing increased bronchopulmonary tract in both hemithoraces and prominence of the pulmonary artery silhouette *(Figure 1A)*.

A transthoracic echocardiogram was performed with the following findings: perimembranous ventricular septal defect of 7 mm, aortic insufficiency jet of 3 mm, fixed fibrous spur type subaortic stenosis (FSSSA) with maximum gradient of 35 mmHg and mean of 19 mmHg, moderate to severe mitral insufficiency with max gradient of 12 mmHg and mean of 19 mmHg (double mitral lesion), right ventricular systolic pressure of 40 mmHg, pulmonary artery systolic pressure of 50 mmHg, left ventricular ejection fraction of 78%, with important left-sided cavities dilatation.

Diagnostic cardiac catheterization was performed, where the following findings were found as follows: pulmonary artery pressure of 100/70 mmHg, mean 87 mmHg; aorta 111/57 mmHg, mean 80 mmHg; right atrium 4/0 mmHg, mean 6 mmHg, maximum peak-to-peak gradient: 5 mmHg; right ventricle 95/25 mmHg, telediastolic 14 mmHg; left ventricle 98/3 mmHg, telediastolic 7 mmHg, maximum peak-to-peak gradient 13 mmHg. Severe pulmonary hypertension and small ventricular septal defect of 7 mm. Initially there was no evidence of left-right shunt in the catheterization. Subsequent analysis allows us to identify it *(Figure 1B)*.

In addition to the previous studies, CT angiography was performed, which was not useful due to poor radiological technique. Therefore, the defect is not evidenced by this imaging study. However, a reconstruction of the CT angiography was requested *(Figure 1C)*.

The patient was scheduled for operation in our service for closure of ventricular septal defect, mitral valve replacement with a mechanical prosthesis and subaortic stenosis resection.

Surgical correction was performed by median sternotomy with extracorporeal circulation. On opening the pericardium, as an operative finding, aortopulmonary communication was observed *(Figure 2A)*. At that time the diagnosis of aortopulmonary window is made in the operating room.

Correction was performed by initiating aortic cannulation with purse-string suture 3-0 polypropylene and superior and inferior vena cava cannulation with purse-string suture 4-0 polypropylene. Subsequently, heparinization with 15,000 IU was performed and cardiopulmonary bypass was started. Double aorta and APW clamping is performed. Cardioplegia is administered in the first half via the aortic root. Second half is administered through left and right ostium until electromechanical arrest.

For resolution of PDA, a pericardial patch is placed. To begin the resolution of the defect, the aortopulmonary window section was performed. The aim of the correction was to achieve complete separation of the pulmonary trunk and the aorta. This separation allowed for the individual repair of each structure. Two pericardial patches were placed to perform the plasty of the involved structures. One of the patches was placed in the aortic mouth of the defect and the second was placed in the pulmonary trunk *(Figure 2B)*. After correction of the aortopulmonary window defect, subaortic stenosis resection was performed.

For valve replacement, a transseptal approach is used. A 25mm mechanical valve was placed in the mitral position. The interatrial septum is closed with prolene 4-0 surjete. Finally, cavity purging and aortic unclamping are performed. We searched for previously reported interventricular septal defect without finding evidence of the defect. The time of extracorporeal circulation was 3 hours. The aortic clamp time was 2 hours and 34 minutes.

The day after surgery, a new control echocardiogram was performed, where the following findings were reported: FSSSA resection in good condition, recording a peak aortic outflow gradient of 8 mmHg and mean of 4 mmHg; mechanical mitral valve is observed in proper position with an opening area of 1.7 cm^2 and a peak gradient of 6 mmHg and mean of 3 mmHg, right ventricle systolic pressure 6 mmHg, pulmonary artery systolic pressure 16 mmHg, left ventricle ejection fraction 62%. Length of stay in the Intensive Care Unit of 3 days. Postoperative course uneventful. In-hospital discharge on the fourth postoperative day.

A postoperative tomography was performed to corroborate the correction of the defect, which reported cardiomegaly, pulmonary arteries at the level of the root of the right pulmonary branch with a decrease in its caliber of 9 mm, adequate course of contrast medium without leaks. Ascending aorta, arch and the descending aorta with adequate diameters and no filling defects were detected *(Figure 3)*.

COMMENT

The presentation of this case description has different situations that are of interest for analysis and review. We present the case of a 17-year-old female patient with an initial diagnosis of fixed spur type subaortic stenosis, moderate to severe mitral insufficiency and 7 mm perimembranous ventricular septal defect. In relation to the literature and the latest classification established in the Congenital Heart Surgery Database and Nomenclature Project: aortopulmonary window, a type II APW associated with ductus arteriosus was presented.[8]

The diagnosis, as has been demonstrated in several case series, is performed by transthoracic two-dimensional echocardiography (ECOTT) with Doppler.[1] Among other utilities of ECOTT is the establishment of the degree of pulmonary hypertension. In the case of our patient, last ECOTT demonstrated a pulmonary artery systolic pressure of 50 mmHg. By non-invasive methods, such as echocardiography, the average in healthy adolescents is a normal range of 20 mmHg.[15] In the cardiac catheterization performed months before surgery, severe pulmonary hypertension was evidenced. In our case, different echocardiograms were performed at different times, and the main abnormality, APW, went unnoticed. Therefore, the diagnosis of APW was made in the operating room. There are other studies that support the diagnosis, such as catheterization and CT angiography. In our case, both studies were performed. The quality of the CT angiographic study was not significant to make the diagnosis. On the other hand, catheterization did not show clear images to make a clear diagnosis. In such a way that it was decided to opt for surgical intervention presenting the diagnoses of FSSSA, severe mitral insufficiency and ventricular septal defect.

In a study conducted in 2019 by El Dick et al.[1] a cohort of 62 adult patients was studied; 41 patients presented type I proximal, 13 type II distal, 8 type III total defects. In the case presented here, we can make the correlation with

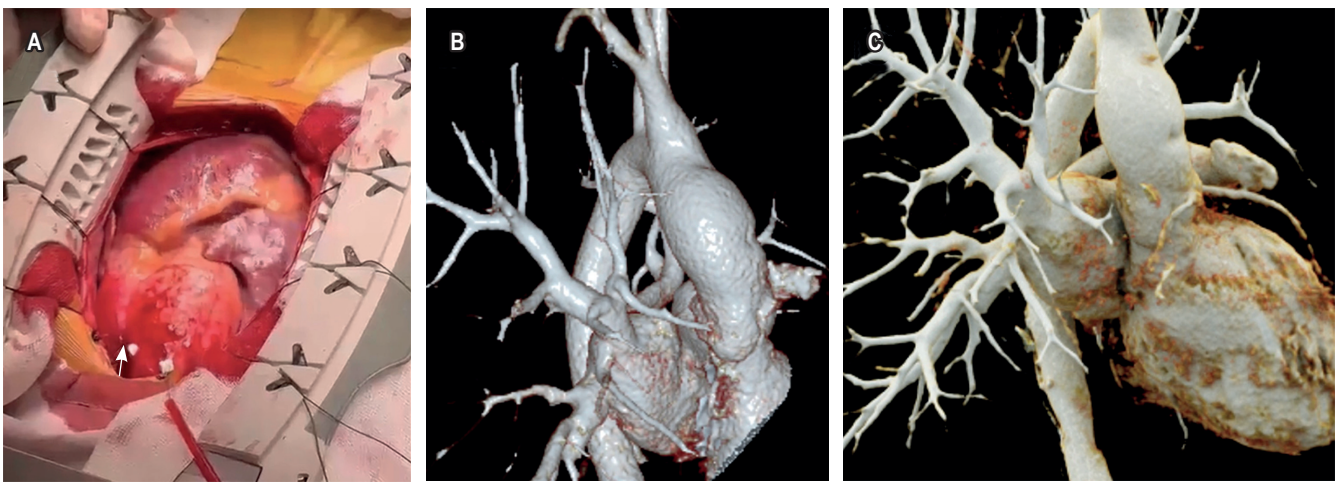

Figure 3: A) Image of the defect when opening the cavity. **B-C)** Antiotomography images after surgery with reconstruction, showing the closure of both vessels without defects.

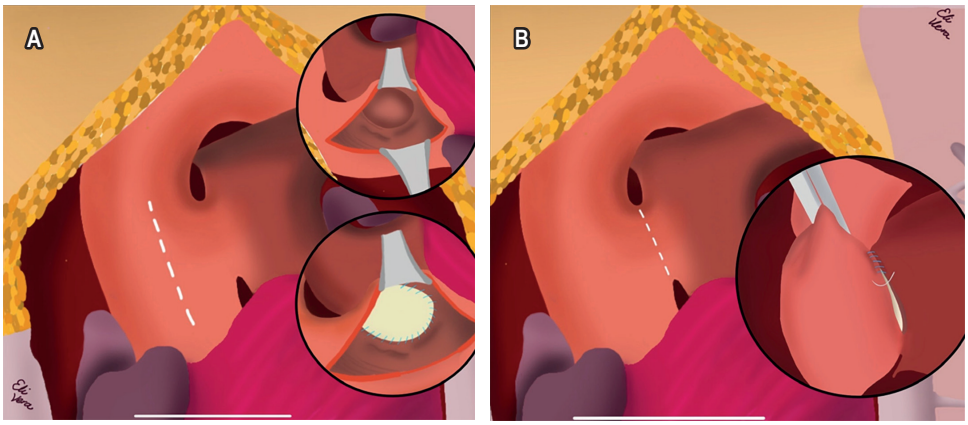

Figure 4:

A) Illustration showing the transaortic approach and suturing of the patch over the defect through the incision. **B)** Illustration showing the transpulmonary approach and sandwich patch suture.

respect to the last classification. Our patient presented a type II or distal defect.

According to a study published in 2020 by Bin-Moallim et al.[9] where 20 patients aged from 4.8 to 5.8 months were analyzed, 2 patients presented simple APW while 18 patients were associated with other malformations. In this study, the most frequently associated defect was atrial septal defect in 40% of the studied population. In relation to the PDA they reported in 40% of the patients. It is worth mentioning that in 2/3 of the patients with PDA they found interruption of the aortic arch. Regarding the complications presented during a follow-up of 4.5 years, 85% of the patients in the cohort studied remained asymptomatic without residual APW and did not require reintervention. Complications occurred in three patients which included mild residual lesions, including mild supra-pulmonary valvular stenosis, leakage through the APW patch and mild pulmonary artery stenosis.[9]

The resolution of the defect can be done through several approaches. The approach conventionally used is the transaortic approach *(Figure 4A)*. It is useful for the variant of the defect where a more extensive part of the aorta is involved. It is useful in cases where the defect extends to the right pulmonary artery. Allows better visualization of the posterior portion of the aorta.[16] The anterior approach consists of vertical opening of the anterior wall of the defect. It is performed using the anterior sandwich patch technique. The inferior, posterior and superior border of the defect is sutured. When the incision is closed, the patch is incorporated into the suture line *(Figure 4B)*. A transpulmonary approach can be performed. This is performed when visualization of the coronary ostia is difficult. It consists of opening the pulmonary artery and placing a patch through it. A pulmonary artery flap can be performed. The latter approach is not routinely recommended due to the risk of reoperation for severe pulmonary artery stenosis.[17]

CONCLUSIONS

The surgical correction in our patient involved a challenge during the intervention because prior to the procedure, the presence of APW was not contemplated. Once surgery was performed, the aorto-pulmonary window was divided by placing two patches, one in each structure (aorta and pulmonary artery), thus avoiding the risk of fistulas or residual communications. One of the complications that our patient presented, as described in some case series, is mild supra-pulmonary stenosis.[9] The technique used in our case has been described as a variant of the anterior approach. The only disadvantage described to this approach is the time that the surgeon will invest in performing it. Generates good benefits for the patient and few or mild complications.[17]

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