CASE REPORT

Pulmonary Atresia with Ventricular Septal Defect, How Far Can We Manage the Patient?

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ABSTRACT

Pulmonary atresia and ventricular septal defect (PA-VSD) with major aorto-pulmonary collaterals (MAPCAs) is a complex and extremely heterogeneous anomaly. Most untreated patients die in their first decade of life because of intractable congestive heart failure or respiratory distress. PA-VSD is characterized by a wide variety of anatomy of central pulmonary artery and nature of collateral lung perfusion. In most patients, collateral perfusion is provided either by MAPCAs or by patent ductus arteriosus (PDA). The management of infants and children with pulmonary atresia, ventricular septal defect, and MAPCAs has proven to be challenging. Therapeutic approaches include one-stage surgical repair, staged unifocalization, shunting, and coiling of collateral vessels. Results have been variable and frustrating. In this case report, we discuss the characteristic, variants, and how far we can manage the patient who suffered from PA-VSD.
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INTRODUCTION

Pulmonary atresia is a rare case, the frequency pulmonary atresia patients with VSD (PA-VSD) is around 2,5-3,4% of all congenital heart malformations. The etiology of PA-VSD is multifactorial, genetic, environmental and family factors. It requires a therapeutic strategy based on catheterization combined with surgical repair to obtain better results in the coming years (1). We report a case of PA-VSD with non-confluent pulmonary artery.

CASE REPORT

A 3-year-old girl came to clinic with chief complaint becoming blue whenever she cried. This happened for almost 2 years. She was told having heart defect since baby. She was born normally with the weight of 2,7 kg.

From the physical examination, blood pressure was 90/70 mmHg, HR 110 x/min regular, RR 24 x/m, with Oxygen saturation was 80-82% and appears cyanotic.

From the thorax examination there was grade III systolic murmur grade III at ICS IV Left Parasternal Line with continuous murmur in ICS II Left Parasternal Line. There are no rales and no wheezing. Her extremities are cyanotic with clubbing fingers.

Laboratory results showed the Hemoglobin level was 14,3 g/dL, hematocrit level was 46%, creatinine 0,4 mg/dL and others were unremarkable.

From the chest X-Ray the cardiac apex was seen tilted upward, pulmonary segment is concave. TTE showed dilated Right Atrium (RA), Hypertrophy Right Ventricle (RV), atretic pulmonal valves with Secundum ASD with size 1,44 cm, Malalignment VSD with size 1,39 cm and Patent Ductus Arteriosus 0,51 cm with arotic overriding 42 %.

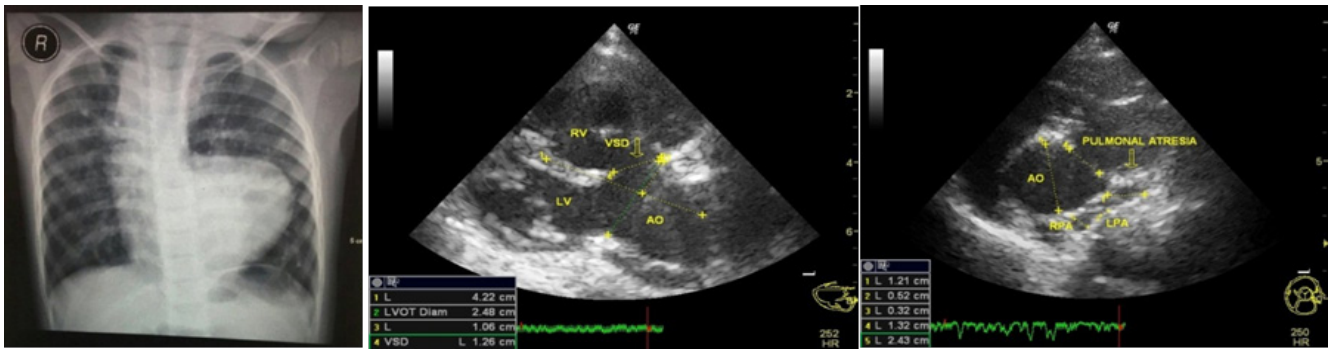

Figure 1: Chest X-Ray and Trans Thoracal Echocardiography (TTE)

From the right ventriculography : there is pulmonary atresia, no flow to the main trunk pulmonary artery. Aortography : contrast fill the right and left pulmonary artery from the Patent ductus arteriosus and MAPCA, the origin of the left pulmonary artery seem narrowed (black arrow). The final result showed Pulmonal Atresia with Large VSD, Large Secundum ASD and Moderate size of PDA.

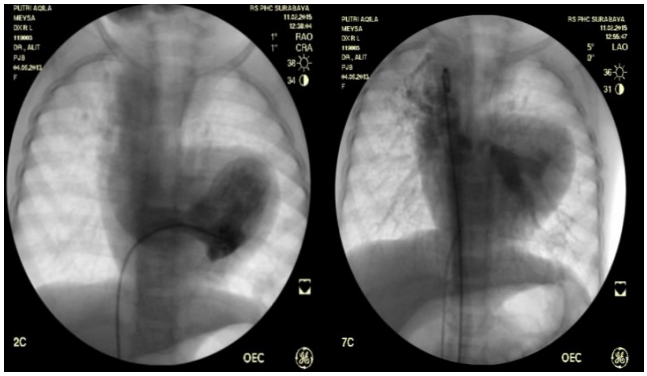

Figure 2: Right ventriculography and aortography

We assess the patient with Pulmonary Atresia with Ventricular Septal Defect, Atrial Septal Defect, Patent Ductus Arteriosus, and Major Aorto-Pulmonary Collateral Arteries. She was treated with supplemental iron syrup due to her hemoglobin level being only 14 g/dL. This case is quite challenging since the girl is already a toddler (3 years old), without any symptoms of hypoxic spells nor heart failure and this pulmonary artery of this patient was non-confluent. The second choice is surgical repair with multiple stages of unifocalization of the pulmonary artery.

DISCUSSION

Terminology and Classification

Pulmonary Atresia with Ventricular Septal Defect (PA-VSD) is a cyanotic congenital disorder with the following characteristics: Under development of the RVOT (Right Ventricle Outflow Tract) in the infundibulum site accompanied with atretic pulmonal valve and there are large VSD with overriding Aorta. Pathologically, PA-VSD is often considered the most severe and final form of Tetralogy of Fallot (TOF)(2).

The Congenital Heart Surgeons Society classifies PA-VSD based on the complexity of the pulmonary blood supply into three types :

1. Type A: There are native pulmonary arteries and confluent, the PDA supply the pulmonary vascular, without collateral arteries.
2. Type B: Both native pulmonary artery and collateral arteries are growing. The lung segments get a double supply. PDA and MAPCAs supply the native pulmonary artery.
3. Type C: There is no native pulmonary artery, the pulmonary blood supply comes from collateral arteries only. (1,2).

Clinical Manifestations and Supporting Measurements

Approximately 65% of patients with atresia pulmonary come to the physician or hospital since baby. Patients who seek for medical treatment in older age usually because their pulmonary blood flow is high enough that the cyanosis appears late. The sign of patient with pulmonary atresia are : cyanosis(50%), heart failure (25%) and murmurs with mild cyanosis, without and with failure to thrive (25%).(1,2) In duct-dependent case, the baby will experience clinical symptoms when the duct begins to narrow. If the duct closes, hypoxia, acidosis, shock occurs and the baby might die immediately.(1)

On physical examination, there was an increased pulsation inferior to the sternum (due to RVH). From auscultation there are grade III/VI systolic murmurs at the Left Lower Sternal Border. Continuous murmurs are heard in the upper chest if there is PDA and if MAPCAs are present, the murmurs will heard diffusely throughout the chest and back.(3) From the Chest-X Ray we will find the shape of the heart is like a boot (coeur en sabot) or looks like a wooden golf club. The MPA segment is small or absent, creating a concave shape beneath the dilated aortic arch. RVH make the apex of the heart to lift upward (from the diaphragm). (1,2,4)

Supporting Measurements

Trans Thoracal Echocardiography has become the gold standard for detecting intracardiac defects but has limitations for describing extracardiac vascular structures. In the PA-VSD the echocardiography

findings are similar to the classic anatomy of TOF but there is without any connection between the RV and the pulmonary artery. From long-axis parasternal position, the aorta is overriding the mal-aligned VSD. There is hypoplastic infundibulum RV (from short axis view). From the suprasternal and high parasternal positions, we can see the size and the confluence of the pulmonary arteries, the aortic arch and assess the patency of the ductus arteriosus. (5)

Cardiac catheterization has capability for measuring hemodynamic and to reveal the things that cannot be obtained from echocardiography such as coronary anatomy, aorto-pulmonary collateral arteries, and visualize the native pulmonary artery (present or absent). Computed Tomography and or Magnetic Resonance angiography provides an alternative modality to conventional angiography for defining the RVOT, MPA, pulmonary artery branches and MAPCAs. (1)

From the echocardiography and cardiac catheterization showed that she suffered from pulmonary atresia with VSD. She has no main pulmonary artery trunk, but her lungs got enough blood supply from PDA and MAPCAs. All the examinations lead to non-confluent PA-VSD. The clinical condition of the patient is quite good because there are MAPCAs and there is PDA that give enough blood supply to the lungs.

Management

Medical treatment

Management of patients with severe cyanosis requires the administration of 100% oxygen immediately, although it is not very effective when PBF decreases rapidly. Prostaglandin (PG) E1 infusion should be started immediately to dilate the ductus arteriosus and increase PBF. If improvement occurs, the decision on the optimal time for surgery can be postponed, but if hypoxia and severe acidosis occur then the decision must be made as soon as possible. (3,5)

Intervention

In PA-VSD, the pulmonary artery may be too small to accommodate sufficient pulmonary blood flow. Recently, the choices are :

1. Angioplasty using a balloon on the pulmonary valve annulus (make the diameter of the pulmonary artery wider).
2. Placement of a balloon and/or stent at the source of the PBF (either collateral artery or duct). The goal is to maintain PDA flow so that the baby survives. (5)

Surgical

Connection between the RV and the main pulmonary artery (PA) should be established as early as possible. Because it will make the pulmonary arteries enlarge rapidly during the first year of life and improve pulmonary artery distribution for alveolar development. The choice for surigcal management are :

1.Central shunt operation. It directly connecting the ascending aorta to the hypoplastic MPA to regenerate the peripheral pulmonary artery. Continue with unifocalization, create RV-PA connection and VSD closure. (5)

2.RV to PA connection with one-stage repair or multiple-stages repair. Primary repair consists of closing the VSD, maintaining the connection between the RV and the pulmonary artery which has been unifocalized (5). Multiple stages repair consist of : Connect the RV to the hypoplastic pulmonary artery (conduit) using homograft then continue to unifocalization procedure. Last step is VSD closure either without or with fenestration in the next 1-3 years. (5)

The patient will likely require complete and staged surgical management. The management for the first case is challenging, whether is it surgical therapy or conservative management by preserving the duct and collaterals. The discussions between the cardiology pediatrician and the thoracic surgeon for surgical correction still on going. There are two choices of management: the first choice is conservative management. We will observe, wait, and see the symptoms of the patient, since the patient is in stable condition.

The surgical management requires congenital heart surgeon who is experienced in performing pulmonary artery unifocalization since there is no normal main pulmonary artery trunk visible (non-confluent). The next stage are to do surgical closure of the shunt (VSD,ASD and PDA), and also closing the MAPCA since the MAPCA might cause over blood flow of the pulmonary artery and induce heart failure.

Prognosis

Many sequelae are possible if complete surgical repair is not performed. PA-VSD patients whose pulmonary blood flow relies on systemic collaterals will experience progressive cyanosis and polycythemia throughout adulthood. Even though complete repair has been performed, gradual decline of conduit function, reduced lumen diameter, calcification and worsening of valve function may occur in several years.(1,3,4)

CONCLUSION

There are a case report of PA-VSD. The patient was in stable and good condition due to the PDA and collaterals that give enough blood and oxygen supply to the lungs. It is challenging to do complete repair of the patients. How far can we manage the patient with PA-VSD is still limited, in terms of human resources, facilities and patient compliance.

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