

CLINICAL CASE

Occlusion of aortopulmonary collateral arteries with Amplatzer® Vascular Plug II device. Case report

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ABSTRACT

Background. Pulmonary atresia with ventricular septal defect (VSD) is a complex heart disease with an incidence of ~2% of all congenital heart diseases. It is frequently associated with 22q11 deletion syndrome. Due to extensive anatomic variability, it is necessary to accurately establish an individualized surgical/medical plan.

Case report. We report the case of a 2-year-old patient with pulmonary atresia and two associated large septal aortopulmonary collaterals. This patient underwent Rastelli surgery at which time it was possible to ligate one of the collateral arteries. The remaining were embolized by transcatheter device with total occlusion.

Conclusions. Management of collateral arteries associated with pulmonary atresia with VSD is complex but is accessible with transcatheter treatment with device implantation and with minimal morbidity.

Key words: transcatheter embolization of collaterals, pulmonary atresia, aortopulmonary collaterals.

INTRODUCTION

Pulmonary atresia with interventricular communication (PA + IVC) can be considered the most serious variety of tetralogy of Fallot, although the anatomy is often more complex. Therefore, it requires a precise anatomic study to establish an appropriate medical and surgical therapeutic program for each patient. PA + IVC has an incidence of 2% among all congenital heart diseases.¹ Anatomic variability includes hypoplasia of the trunk and pulmonary arteries, pulmonary vascular abnormalities and the presence of abnormal flow coming, in most cases, from the descending aorta to the great vessels, generally between 1 and 8.^{2,3} They provide perfusion to various portions of both lungs and may contribute to increased pulmonary

flow. Surgical access for ligation or section of these collaterals can be complicated. Transcatheter procedures for occlusion of those abnormal arteries can be carried out, particularly if they are clinically significant.

The goal of the surgery should be to connect the right ventricle to the pulmonary artery and, if any collateral arteries exist, to connect as many of these (targeting) to the central pulmonary circulation or if the connection to the right ventricle-pulmonary artery is sufficient, collateral circulation can be ruled out by surgery or embolism. Since 2000, the *Congenital Heart Surgeons Society* published the characterization of pulmonary circulation based on angiographic findings, which provides therapeutic guidance according to the individual's anatomy and physiology.⁴

It is worth noting that PA + IVC are strongly associated with 22q11 deletion syndrome, particularly when the great collaterals are present.⁵ Therefore, the characteristic facies must be searched for as well as chromosomal study because the presence of this syndrome has major implications for the future of the children.

We present the case of a patient with PA + IVC who was operated on by connecting the pulmonary ventricle with a prosthetic graft. Subsequently, an occlusion was

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conducted of a large collateral artery of the descending aorta to the pulmonary circulation using an Amplatzer® Vascular Plug II.

CLINICAL CASE

We present the case of a 3-year-old preschooler. He is the third child in his family and the product of a normal pregnancy resulting in a vaginal delivery and weighing 2690 g, 52 cm in length and Apgar of 8-9. There was no relevant family history. Perioral and ungual cyanosis was detected that increased from the first month of life. There was respiratory distress associated with intercostal retractions. The patient was referred to our institution at 10 months of age. On admission no abnormal facies were found. There was grade II cyanosis, left precordial hyperactivity and continuous murmurs in the periphery of both lung fields with II/VI intensity. The second sound was unique. Imaging studies were performed to rule out the 22q11 deletion and reported pulmonary atresia with interventricular communication, with a thrust of 50%, infundibular masses obstructing the right ventricular outflow tract and valvular atresia with severe hypoplastic pulmonary annulus. Pulmonary arteries were confluent: the right was 9.68 mm in diameter ($Z = +2$) and the left 9.49 mm ($Z = +1$). The first cardiac catheterization performed at 1 year of age showed two large collaterals. One began from the right subclavian and communicated with both pulmonary artery branches (Figure 1). The other collateral emerged from the descending aorta (5-mm diameter) and with communicating flow directly to the right branch of the pulmonary artery. The pressure in these two systems was similar at 44/26 mmHg (mean 34 mmHg). With these findings, we decided to carry out a total correction at 2 years of age. During surgery, data reported in the cardiac catheterization were confirmed. A patch was placed to close the interventricular communication, ligation of the collateral dependent of the right subclavian was done and a Hancock® valved tube of 16 mm in diameter was implanted between the right ventricular outflow tract and pulmonary artery without complications. Subsequent progress was satisfactory. However, the patient persisted with mild tachypnea in the months following surgery. Nine months later, during a follow-up consultation, tachypnea was confirmed. A control echocardiogram revealed a significant flow in the descending

aorta collateral. A diagnostic cardiac catheterization was scheduled to evaluate the possibility of occlusion.

The second catheterization showed an increase in saturation of 15% (66% in vena cava to 81% in the right branch of the pulmonary artery) with pressure of 30/10 mmHg (mean 12 mmHg) in the right branch of the pulmonary artery. The discharge ratio was 1.8:1 caused by the communicating collateral artery. The maximum diameter of this was 4.8 mm, which prompted the decision for occlusion with an 8-mm Amplatzer® Vascular Plug II (Figure 2). Immediately after the occlusion, saturation in the right branch was normalized. The pressure in this branch decreased to 17/7 mmHg (mean 10 mmHg) and the discharge ratio stabilized at 1:1. There were no complications during the procedure and the patient was discharged the same day.

DISCUSSION

Large-size aortopulmonary collateral arteries that connect the systemic circulation with the pulmonary circulation are a common finding in PA + IVC and represent the remainder of embryonic ventral splanchnic arteries which, under normal conditions, must demonstrate a regression as the pulmonary arterial circulation is established during the first weeks of gestation.⁶ However, these arteries become collateral arteries with various sizes and functionality with direct relation to poorly developed pulmonary arterial circulation, specifically in cases of pulmonary atresia. This results in a wide range of pulmonary circulatory disorders ranging from simple confluent branch pulmonary atresia, hypoplastic of pulmonary arteries, loss of confluence of the pulmonary arteries, presence of major aortopulmonary collaterals that may or may not be communicating with the pulmonary flow to agenesis of true pulmonary branches. It is clear that each of the possible anatomic variants needs to be individually designed for surgical correction, whose main goal is to restore, as far as possible, the ventricle-pulmonary circulation. However, this will depend on the anatomic characteristics of the pulmonary and collateral arteries. The options, thus far, are to restore pulmonary ventricle continuity through a patch or valved conduit (according to the intracardiac anatomy), total focalization or in stages (according to the anatomy of the pulmonary and collateral arteries) or decide whether or not to close the

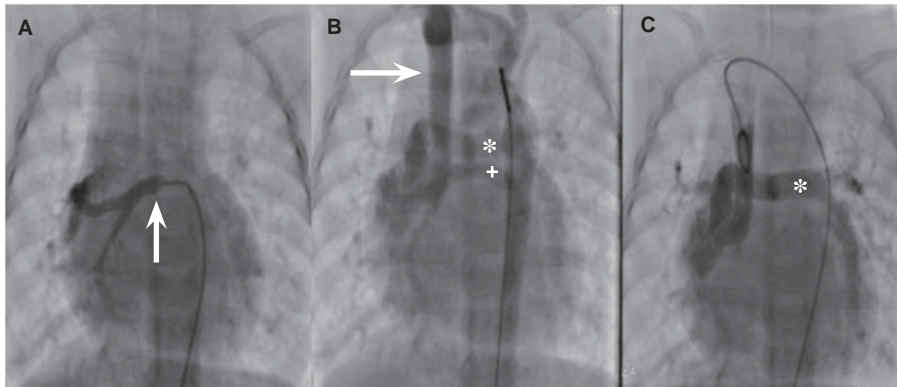


Figure 1. (A) Catheter located in the communicating large collateral artery (arrow) arising from the descending aorta to the right branch of the pulmonary artery. (B) Aortic angiography showing the collateral artery, which originates from the right subclavian artery (arrow) and connects to the right branch of the pulmonary artery. Note the confluence of the true pulmonary arteries (*) and collateral artery from the descending aorta (+). (C) Cannulation the collateral artery showing the communication towards the true branches of the pulmonary artery (*).

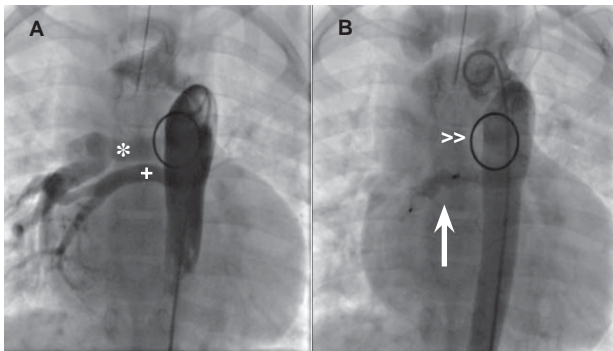


Figure 2. (A) Angiography in descending aorta shows the large collateral artery (+) communicating with the right branch of the pulmonary artery (*) with a diameter is 4.8 mm. (B) Arrow indicates the Amplatzer Vascular Plug II device, totally occluding the collateral artery circulation. >>Pulmonary valve prosthesis.

interventricular communication (according to hypoplasia of the pulmonary branches).⁷⁻⁹

In this case, ventricular anatomy and pulmonary branches allowed a connection from the right ventricle to the pulmonary artery where a valved conduit was implanted. During the surgical procedure, it was possible to link the large right subclavian collateral dependently. However, it was difficult to surgically remove the collateral that derived from the descending aorta. During follow-up it was evident that pulmonary hyperflow was generated because of a QP/QS ratio of 1.8:1 and 15% increase in saturation between the superior vena cava and right pulmonary artery. Therefore, a transcatheter closure of the collateral was decided upon. Due to a diameter of 4.8 mm, it was considered that the best option was to use an 8-mm Amplatzer® Vascular Plug II allowing complete and immedia-

te closure without complications of this collateral, normalizing the pulmonary circulation.

Patients with PA + IVC may have multiple and large collateral arteries without any obstruction, as in this case, producing an excessive amount of blood to the lungs and predisposing to heart failure. Therefore, it is necessary to intervene in order to suppress this circulation. The traditional method is collateral ligation. However, on occasion, alternative transcatheter treatment is useful in view of difficult or remote sites of the operative field. Historically, various materials have been used for embolization including adhesive tissues such as Gelfoam® (purified porcine skin gelatin), silicon balls and Gianturco coils.¹⁰ At present it is possible that these are the most widely used materials. They have been reported as successful in >90% of the cases.⁶ However, in some instances, due to the position of the vessels, diameter and their own tortuosity, the implant may be difficult or require several spirals.

The development of new instruments such as the Amplatzer® Vascular Plug (AVP) device made with a nickel-titanium (nitinol) mesh with self-expandable memory¹¹ has allowed the simplification of the occlusion of selected cases of various arteriovenous malformations, including the large collaterals related to PA + IVC. The current version of AVP is the II model, which was approved by the FDA in 2007 for clinical use.¹² This new version consists of three segments with six dense layers of nitinol, which provide better occlusive properties. Furthermore, the implant device is simple with low profile catheters. Furthermore, the most worrisome complication, which is migration of the device, is minimized because it is a self-expanding device that generates sufficient radial force to remain stationary once deployed. In cases of PA + IVC

with large aortopulmonary collaterals, a close relationship exists with the 22q11 deletion syndrome. There is some clinical evidence that relates the collaterals that arise from the brachiocephalic vessels with this syndrome at higher rates than those with large collateral that arise directly from the descending aorta.¹³ This information is important, particularly for the natural history of patients with 22q11 deletion, not only from a cardiological sense but also from a neurological and psychiatric perspective. In this case, this possibility was ruled out by a fluorescence in situ hybridization study (FISH). It is important to mention that the long-term evolution of the collaterals is uncertain and may influence pulmonary vascular disease in some cases or also gradually evolve to stenosis with progressive lung damage. For this reason, reestablishment of the pulmonary circulation in the most possible physiological manner is mandatory in PA + IVC.

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