CASE REPORT

Isolated persistence of the fifth aortic arch in an infant presenting with congestive heart failure

Giovanni Meliota[1], Maristella Lombardi[1], Pierluigi Zaza[1], Maria Rosaria Tagliente[1], Paolo Versacci[2], Gabriele Scalzo[1], Ugo Vairo[1]

[1]Department of Pediatric Cardiology, Giovanni XXIII Pediatric Hospital, Bari, Italy, [2]Department of Peditarics, Obstetrics and Gynecology, Sapienza - University of Rome, Rome, Italy

ABSTRACT

The persistence of the fifth aortic arch (PFAA) in postnatal life is an extremely rare and controversial cardiovascular malformation. PFAA is defined as an extra-pericardial vessel arising from the ascending aorta proximal to the origin of the brachiocephalic arteries, terminating either in the dorsal aorta or in the pulmonary arteries through the persistently patent arterial duct. An isolated PFAA with systemic-to-pulmonary connection best fits this definition, while the vast majority of cases reported as PFAA may have alternative embryological explanations. We present a unique case of a 5-week-old patient with an isolated PFAA with systemic-to-pulmonary connection, who presented with congestive heart failure. A first differential diagnosis was made with distal aortopulmonary window and an atypical patent arterial duct. A careful analysis of the case and a systematic review of the literature made us conclude for an isolated PFAA, which is one of the only five cases ever reported.

Keywords: Aortic arch anomalies, aortopulmonary window, congestive heart failure, persistence of fifth aortic arch

INTRODUCTION

The persistence of the fifth aortic arch (PFAA) in postnatal life is a cardiovascular malformation so uncommon in humans that its definition and true embryological derivation are still a subject of controversy.[1] In 1969, van Praagh and van Praagh[2] were the first to suggest that the so-called double-lumen aorta could be interpreted as the persistence of the artery of the fifth aortic arch. Since then, the PFAA has been anecdotally reported in literature with variable anatomical forms. According to a retrospective analysis of over 2000 cardio-pathological specimens, its incidence is about 1 in every 330 autopsy cases.[3]

PFAA is frequently described in association with complex cardiac heart defects and its presence is usually beneficial to the associated cardiac anomalies.[4] Freedom *et al.*[5] defined four different subtypes based on the anatomical and physiological characteristics of the existing case reports[4] [Table 1 and Figure 1].

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CLINICAL SUMMARY

A 5-week-old male patient was referred to our department due to a suspected aortic arch anomaly. He was delivered through spontaneous vaginal birth at full-term after a normal course of pregnancy, with a birth weight of 3.07 kg. After the first 3 weeks of life, he progressively developed poor weight gain and respiratory distress during feeding. He was then examined by an outpatient pediatric cardiologist and referred to our evaluation.

On hospital admission, the patient was fussy and tachypneic. His body weight was 3.65 kg (<3rd centile).

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Address for correspondence: Dr. Giovanni Meliota, Department of Pediatric Cardiology, Giovanni XXIII Paediatric Hospital, Via Amendola 207, 70126 Bari, Italy. E-mail: giovanni.meliota@gmail.com

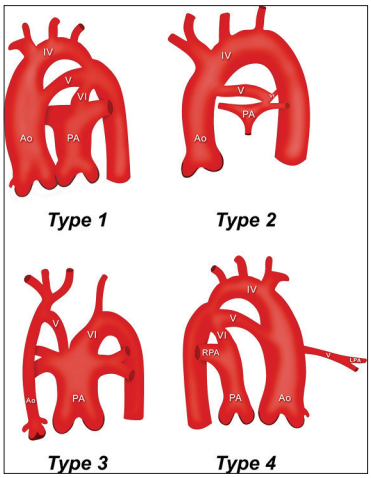

Figure 1: Freedom's classification of PFAA. Top left: Type 1, systemic-to-systemic PFAA. A case of double lumen aortic arch is shown. Top right: Type 2, systemic-to-pulmonary PFAA, in a case associated with pulmonary atresia. Bottom left: Type 3, pulmonary-to-systemic PFAA associated with left-sided obstructive lesions. A case of PFAA associated with aortic atresia and Type B aortic arch interruption. Bottom right: Type 4, Bilateral PFAA, with right double lumen aortic arch and left PFAA. PFAA: Persistence of fifth aortic arch

His heart rate was 170 beats/min, and peripheral oxygen saturation on room air was 99%. Right arm blood pressure was 80/40 mmHg, equal to lower limb blood pressure. He presented no dysmorphic features at general inspection. Physical examination revealed a Grade III continuous machinery murmur, loudest in systole and best heard at the left infraclavicular area and a fine wheezing at thoracic auscultation. The liver was mildly enlarged. Femoral pulses were well palpable.

EKG demonstrated sinus rhythm with signs of left ventricular hypertrophy. Chest X-ray displayed cardiomegaly with increased pulmonary vascular markings.

Echocardiography revealed normal sequential anatomy with no intra-cardiac defects. Signs of left-sided volume overload were detected with a functional moderate mitral regurgitation. Global biventricular systolic function was preserved. An abnormal and large vascular connection was identified between the inferior aspect of the aortic arch and pulmonary artery, causing a large left-to-right shunt [Figure 2]. Given this unusual aortic arch anomaly, regarded as a distal aortopulmonary window or a large atypical patent arterial duct, it was decided to further investigate the anatomy with cardiac catheterization.

Aortography showed an extremely large and short vascular structure arising from the inferior concave surface of the aortic arch, beneath the origin of the brachiocephalic artery, and connected to the superior margin of the bifurcation of the pulmonary trunk, causing a systemic-to-pulmonary shunt [Figure 3]. It was recorded a total oxygen saturation step up of 20%

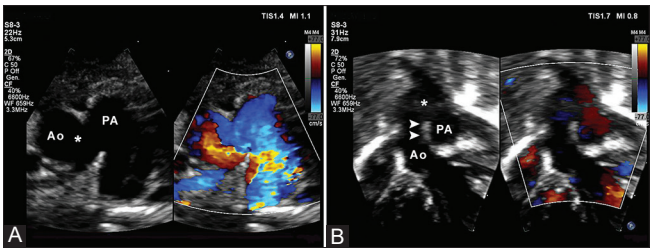

Figure 2: Echocardiographic assessment. Panel A. From a modified parasternal short-axis view, a large and short connection (asterisk) is highlighted between ascending aorta and distal pulmonary artery, just proximal to bifurcation, resembling a distal aorto-pulmonary window. Panel B. Subxiphoid long axis view confirms the presence of a large connection (asterisk) between ascending aorta and pulmonary trunk, but points out a well developed aorto-pulmonary septum (arrow-head)

Table 1: Freedom's classification of persistence of fifth aortic arch, modified from Lloyd *et al.*

Classification	Type of connection	Possible presentation
Type 1	Systemic-to-systemic	Double lumen aortic arch
Type 2	Systemic-to-pulmonary	Isolated or associated with the right-sided obstructive lesion
Type 3	Pulmonary-to-systemic	Associated with left-sided obstructive lesions
Type 4	Bilateral	One case of right double-lumen aortic arch and left PFAA

PFAA: Persistence of fifth aortic arch

from the right atrium to the pulmonary artery and equal aortic and pulmonary artery pressures.

The patient was stabilized in a few days with intravenous furosemide and oral captopril therapy and scheduled for elective surgical intervention. The operation was performed through a median sternotomy. An extrapericardial duct-like vessel with a diameter equal to the ascending aorta was found arising from the ascending aorta, opposite to the origin of the brachiocephalic artery, and terminating on the superior margin of the bifurcation of the pulmonary trunk. Neither arterial duct nor ligament was identifiable in their usual position. The vessel was efficaciously ligated. Intraoperative epicardial echocardiography excluded any significant obstructions of the pulmonary arteries and on the aortic side of the ligated vessel. The postoperative course was uneventful, and the patient has been doing well during the 8 months follow-up period.

DISCUSSION

After a careful analysis of the case and a systematic review of the literature, we concluded that the encountered abnormality was an isolated PFAA with systemic-to-pulmonary connection. It fulfilled the definition criteria proposed by Gupta *et al.*[1] Based on studies on murine and human embryos, he defined a

persistent fifth aortic arch artery as an extra-pericardial vessel arising from the ascending aorta proximal to the origin of the brachiocephalic arteries, terminating either in the dorsal aorta or in the pulmonary arteries through the persistently patent arterial duct.

The possible differential diagnosis could be distal aortopulmonary window or an atypical patent arterial duct. Indeed, the PFAA might resemble a distal aortopulmonary window, as it connects the distal ascending aorta with the bifurcation of the pulmonary trunk. However, while the aortopulmonary windows by definition lie within the pericardial sac, the PFAA is an extrapericardial structure, as the series of paired arch arteries that connect the aortic sac to the initially paired dorsal aortas (and thus the fifth aortic arch artery) are located extrapericardially within the pharyngeal mesenchyme. In our case, the vascular connection was extrapericardial at direct surgical inspection, leading us to exclude the diagnosis of possible distal aortopulmonary window. Moreover, we rejected the suspicion of a patent arterial duct, as in this case the aortic extremity would have arisen beyond the origin of the brachiocephalic artery. Although the absence of any ligamentous connection to descending aorta and the absence of a separate duct or ligamentum make it difficult to rule out a proximal origin of the arterial duct, the abnormal vessel clearly arose from the ascending aorta at direct surgical inspection.

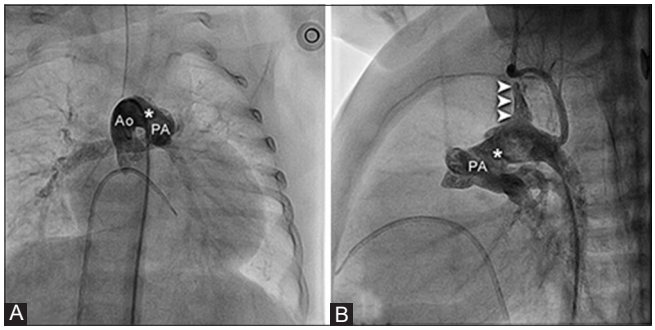

Figure 3: Cardiac catheterization. Panel A. Anterior-posterior projection. A short and large vessel (asterisk) arises proximal to brachiocephalic artery (which is not filled in by contrast, yet) and connects ascending aorta and pulmonary trunk, leading to a significant left-to right shunt. Panel B. Aortography in lateral projection. Contrast flows directly in the abnormal vascular connection (asterisk), which originates beneath brachiocephalic artery (arrow-heads) and has anterior and inferior course to pulmonary artery

Systemic-to-pulmonary PFAA might be associated with critical right-sided obstructive lesions, such as pulmonary atresia and intact ventricular septum, pulmonary atresia and ventricular septal defect, tetralogy of Fallot, and isolated left pulmonary artery.[6] In such cases, PFAA can act as a vital source of pulmonary flow, and some authors reported its responsiveness to intravenous prostaglandin infusion.[7]

Systemic-to-pulmonary PFAA can also be an isolated pathological entity, causing excessive pulmonary blood flow with pulmonary hypertension. Systemic-to-pulmonary PFAA might have been underrecognized in the past because of the similarities in the clinical presentation of the isolated patent arterial duct and the relatively recent publication of a few reports describing this rare entity.[8-10] Our case is one of only five reported cases of isolated systemic-to-pulmonary PFAA in literature [Table 2].

Advances in the field of cardiac imaging have resulted in many cases being reported as PFAA to explain double-lumen aorta, systemic-to-pulmonary arterial channels, abnormal branching of the brachiocephalic arterial structures. However, most of the vascular malformations attributed to the persistence of the fifth aortic arch artery might depend on alternative developmental aberrations.[6] Bamforth *et al.*[11] observed collateral channels extending between the distal insertions of the fourth and sixth arch arteries in up to half of all developing mice, and in developing human embryos too. Such channels have been described as "fifth arch arteries" by other investigators, but they do not originate from the aortic sac and do not run in parallel with the previously existing bilateral channels.[11] In light of the above, many of the reported cases of PFAA are equally well explained on the basis of persistence and abnormal arterial duct remolding, or persistence of those collateral channels. Some of the cases included in Freedom's classification as persistent arteries of the fifth pharyngeal arch do not clearly follow the definition offered by Gupta *et al*. The association of PFAA and left obstructive lesions (Type 3) and bilateral PFAA (Type 4) have been contested[1,4,6] and could have alternative embryological explanations. According to Gupta *et al.*,[6] none of the reported cases of double lumen aorta (Type 1) unequivocally represents the fifth aortic arch artery, but all can be better explained invoking the dorsal collateral channels.

Table 2: All cases of isolated systemic-to-pulmonary persistence of fifth aortic arch reported in literature

Case	Age	Imaging	Treatment	Outcome	Reference
1	double-lumen years	TTE, cath, MRI	Surgery, Dacron patch	Asymptomatic	Chiu *et al.*[8]
2	1 day	TTE, cath	No treatment	Dead at 3 months	Chiu *et al.*[8]
3	2 months	TTE, CT, cath	Surgery, ligation	Asymptomatic	Hwang *et al.*[9]
4	21 wga	Fetal echo, TTE, CT	Surgery	Asymptomatic	Jowett *et al.*[10]
5	5 weeks	TTE, cath	Surgery, ligation	Asymptomatic	Meliota G*

*Present case report. Cath: Cardiac catheterization, CT: Computed tomography, MRI: Magnetic resonance imaging, TTE: Transthoracic echocardiography, wga: Weeks of gestational age

On the other hand, the only human embryo that showed a structure resembling a true fifth aortic arch artery presented a vascular channel originating from the aortic sac, occupying a discrete component of the pharyngeal mesenchyme and terminating on the artery of the sixth.[9] Therefore, the occurrence of an extrapericardial vascular connection between the ascending aorta and the bifurcation of the pulmonary trunk is the most suitable definition of a persistent fifth aortic arch among other candidates.

PFAA is a rare and controversial condition, often misdiagnosed or ignored. An appropriate description of cardiovascular lesions is important in terms of classification and often affects the therapeutic approaches. The aim of our report is to raise awareness of this rare and complex entity, whose presence has to be taken into account when dealing with aortic arch and great vessel abnormalities.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Gupta SK, Gulati GS, Anderson RH. Clarifying the anatomy of the fifth arch artery. Ann Pediatr Cardiol 2016;9:62-7.
2. van Praagh R, van Praagh S. Persistent fifth arterial arch in man. Congenital double-lumen aortic arch. Am J Cardiol 1969;24:279-82.
3. Gerlis LM, Ho SY, Anderson RH, Da Costa P. Persistent 5th aortic arch – A great pretender: Three new covert cases. Int J Cardiol 1989;23:239-47.
4. Lloyd DF, Ho SY, Pushparajah K, Chakraborty S, Nasser M, Uemura H, *et al.* Persistent fifth aortic arch: The "great pretender" in clinical practice. Cardiol Young 2018;28:175-81.
5. Freedom RM, Yoo SJ, Mikailian H, Williams WG. The Natural and Modified History of Congenital Heart Disease. New YorK, NY: Wiley-Blackwell; 2008.
6. Gupta SK, Bamforth SD, Anderson RH. How frequent is the fifth arch artery? Cardiol Young 2015;25:628-46.
7. Zartner P, Schneider MB, Bein G. Prostaglandin E1 sensitive persistent fifth aortic arch type 2. Heart 2000;84:142.
8. Chiu CC, Wu JR, Chen HM, Lin YT. Persistent fifth aortic arch: An ignored and underestimated disease. Jpn Heart J 2000;41:665-71.
9. Hwang MS, Chang YS, Chu JJ, Su WJ. Isolated persistent fifth aortic arch with systemic-to-pulmonary arterial connection. J Thorac Cardiovasc Surg 2003;126:1643-4.
10. Jowett V, Rubens M, Ho SY, Uemura H, Gardiner HM. Prenatal visualization of persistent 5th aortic arch artery. JACC Cardiovasc Imaging 2012;5:1288-9.
11. Bamforth SD, Chaudhry B, Bennett M, Wilson R, Mohun TJ, Van Mierop LH, *et al.* Clarification of the identity of the mammalian fifth pharyngeal arch artery. Clin Anat 2013;26:173-82.