



Uncommon Coexistence of Aortopulmonary Window With Interrupted Bovine Aortic Arch and Patent Ductus Arteriosus in An Adult: A Case Report and Review of Literature

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Abstract

Aortopulmonary window is an opening between the aorta and the pulmonary trunk. Interrupted aortic arch is defined as a loss of luminal continuity between the ascending and descending portions of the aorta. Bovine aortic arch is the most common variant of aortic arch and occurs when the brachiocephalic shares a common origin with the left common carotid artery. The rarity in our case lies in the co-existence of these rare congenital cardiac anomalies with a delayed age of presentation i.e. 27 years which was diagnosed entirely by MDCT and Echocardiography and to the best of our knowledge this co-existence has not been reported so far.

INTRODUCTION

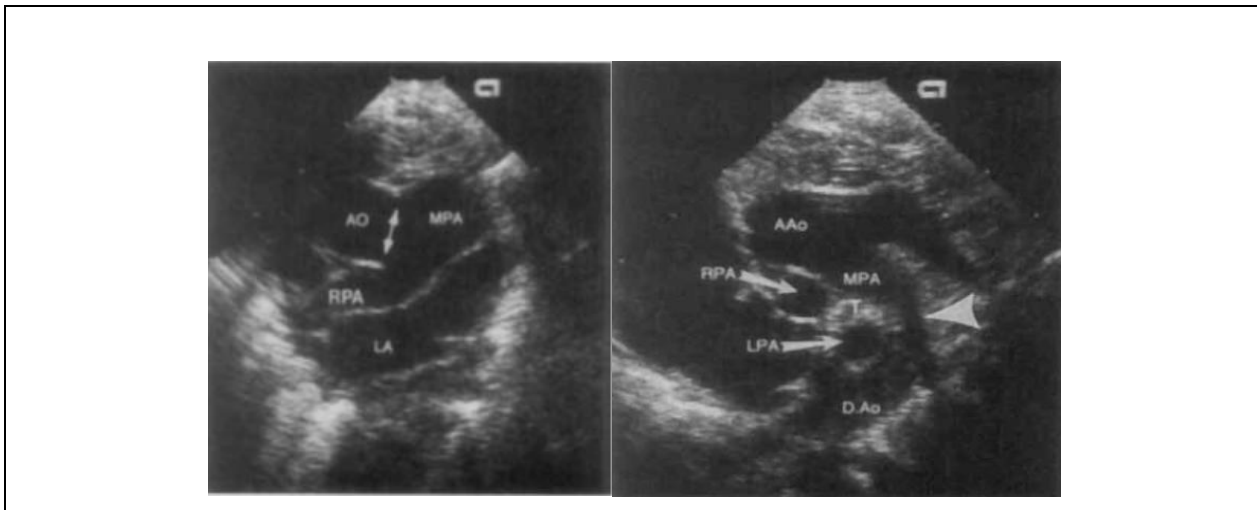
Aortopulmonary window is a rare lesion and its prevalence among patients with congenital heart disease is only 0.1% to 0.2%. The defect is thought to result from incomplete fusion of the truncal ridges, with a resultant persistent communication between the ascending aorta and main pulmonary artery. Currently, approximately 300 cases are cited in the literature about aortopulmonary window. It can also be associated

with other cardiac abnormalities in one-third to one-half of cases. The most common associated lesions are arch abnormalities, specifically interrupted aortic arch, and coarctation of the aorta, Tetralogy of Fallot and abnormal origin of the coronary arteries. Decreased exercise tolerance, and shortness of breath on exertion are the most common symptoms. It is usually diagnosed and surgically treated in childhood.

CASE REPORT

A 27 year old female presented with complaint of exertional dyspnea and syncope episodes since childhood in the cardiology department. General examination revealed bounding pulse, precordial impulse and wide pulse pressure. On cardiac examination a systolic murmur was heard along the left sternal border. Chest x-rays revealed cardiomegaly, narrow vascular pedicle, increased pulmonary notch and increased pulmonary vascular markings consistent with increased pulmonary blood flow. Provisional diagnosis of

aorto-pulmonary communication, tricuspid regurgitation with changes of pulmonary hypertension was made on echocardiography. CT angiography was ordered for confirmation of the diagnosis and the ancillary findings. On subsequent follow up it was informed by the cardiologist, that the patient's condition deteriorated significantly and was showing cyanotic changes possibly due to reversal of persistent ductus arteriosus (Eisenmenger's syndrome)



Imaging Findings

Echocardiography.(Fig-1) Coronal View. Double-Headed Arrow Shows The Aortopulmonary Window Connecting The Ascending Aorta (AO) With The Main Pulmonary Artery (MPA)

(Fig-2) Sagittal View. Arrow Shows The Main Pulmonary Artery (MPA) And Descending Aorta (D.AO) Connected By The Patent Ductus Arteriosus.



FIG-3



FIG-4

Fig-3,4- Axial CT Image Showing Communication Between Ascending Aorta And Dilated Main Pulmonary Artery

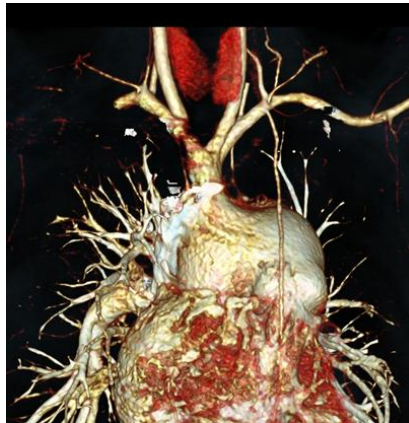


FIG-5



FIG-6

(Fig-5 CT Volume Rendered Reconstruction Image Showing A Large Communication Between Ascending Aorta And Dilated Pulmonary Artery Along With Interrupted Bovine Aortic Arch.

(Fig-6) Patent Ductus Arteriosus Connecting Descending Thoracic Aorta And Dilated Main Pulmonary Artery

COMPUTED TOMOGRAPHY (CT) ANGIOGRAPHY obtained during patient stay in the hospital revealed normal separate origin of proximal ascending aorta (31mm) and pulmonary trunk, large communication (measuring 39 mm) between ascending aorta and the dilated main pulmonary artery (Type 2 Aortopulmonary Window) with common origin of right brachiocephalic artery, left common carotid and subclavian artery from proximal arch of aorta

(9mm) (Bovine Aortic Arch). There is non separate visualization of arch of aorta distal to It (Interrupted Aortic Arch Type A). Descending thoracic aorta is showing communication with the main pulmonary trunk via patent ductus arteriosus

DISCUSSION

Developmentally, the Aortopulmonary window results from incomplete separation of the common tube of the truncus arteriosus and the

aorticopulmonary trunk.¹ During early embryonic development, the aorta and pulmonary arteries separate by growth of a spiral septum dividing the common trunk into the aorta and the pulmonary artery. Incomplete development of these septa results in aortopulmonary septal defect.

This defect may be found anywhere from just above the semilunar valves to the more distal ascending aorta and main pulmonary artery.

Currently, approximately 300 cases are cited in the literature about aortopulmonary window². This defect is present as an isolated lesion in about one half of patients and in conjunction with another defect or more complex heart disease such as arch abnormalities, specifically interrupted aortic arch, and coarctation of the aorta³ in the other half of patients. Abnormal origin of the coronary arteries is commonly associated with aortopulmonary window. Patent ductus arteriosus (PDA) is encountered in almost three fourths of patients with aortopulmonary septal defect^{4,5}

Despite the rarity of aortopulmonary window, the condition has been classified into 3 types by van mierop⁶:

- proximal, or type I (most common);
- distal, or type II; and
- total, or type III.

It has been suggested that, when associated with interrupted aortic arch, aortopulmonary windows are larger with greater distal extension

An interrupted aortic arch (IAA) is an uncommon congenital cardiovascular anomaly where there is a separation between the ascending and descending aorta⁷. It can either be complete or connected by a remnant fibrous band⁸. An

accompanying large ventricular septal defect (VSD) and / or patent ductus arteriosus (PDA) is frequently present. It may account for ~ 1.5% of congenital cardiac anomalies. The defect probably results from regression of both the left and right embryonic aortic arches.

It can be classified to three types according to location of occurrence

- **type a** - second commonest: interruption occurs distal to the left subclavian arterial origin
- **type b** - commonest (> 50 %) : interruption occurs between left common carotid arterial and left subclavian origins
- **type c** - rare : interruption occurs proximal to left common carotid arterial origin

The clinical presentation of aortopulmonary septal defect (APSD) depends on the size of the defect, pulmonary vascular resistance (PVR), and associated anomalies.

In a large defect with falling PVR, aortopulmonary septal defect presents with typical signs and symptoms of congestive heart failure (CHF) with dyspnea and hepatomegaly indistinguishable from those of a large ventricular septal defect (VSD) or ductus arteriosus during first year of life or later in childhood. Pulmonary hypertension can be fatal in almost all cases if not treated early in childhood or adolescence; the few surviving adult patients have symptoms associated with severe pulmonary hypertension, making these cases inoperable.

DIAGNOSIS

The diagnosis of aortopulmonary window and interrupted aortic arch is generally based on medical history, physical examination findings, Chest X rays, Echocardiography, CT angiography and MRI.

Chest x-ray findings reveal cardiomegaly and increased pulmonary vascular markings consistent with increased pulmonary blood flow. Echocardiography is a good tool to establish the diagnosis, to evaluate the presence of associated pulmonary hypertension, the location and size of the communication, as well as associated anomalies. Cardiac catheterization is rarely indicated and reserved for patients who present after early infancy and, therefore, are at risk for elevated pulmonary vascular resistance, or any patient in whom the anatomy cannot be adequately defined by echocardiography⁹. Computed tomography clearly demonstrated the location, type and size of the window. In addition it also demonstrated the other associated anomalies and changes in the lungs. MRI permits evaluation of the direction and quantification of the flow and the presence of other associated anomalies¹⁰. Aortopulmonary window should be considered in the differential diagnosis of a left to right shunt such as large patent ductus arteriosus or ventricular septal defect or a truncus arteriosus. In contrast to truncus arteriosus, both aortic and pulmonary valves are present and normally related

CONCLUSION

Aortopulmonary window with interrupted, bovine aortic arch with patent ductus arteriosus are rare

associated congenital anomalies.. CT angiography and echocardiography are useful noninvasive techniques that can accurately demonstrate the defect and signs of pulmonary hypertension.

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