Interrupted Aortic Arch Associated with Complex Congenital Heart Disease

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Abstract

Interrupted aortic arch (IAA) is a rare congenital malformation of the aortic arch, which occurs in 3 per million live births. This anomaly is defined as a loss of luminal continuity between the ascending and descending portions of the aorta which entails a very poor prognosis without surgical treatment. In most cases, IAA is associated with intracardiac malformations such as VSD, PDA, bicuspid aortic valve, left ventricular outflow tract obstruction or aortopulmonary window. We report a 16-year-old boy with IAA associated with double inlet left ventricle, aneurysmal dilatation of pulmonary artery, giant PDA and severe pulmonary hypertension. To our knowledge, there is no previous report of IAA associated with complex congenital heart disease such as that seen in our case ***(Iranian Heart Journal 2007; 8 (3): 60-63).***

Key words: interrupted aortic arch ■ complex congenital heart disease

Case report

A 16-year-old boy presented with cyanosis, dyspnea on exertion and easy fatiguability since childhood. He had a previous history of cardiac catheterization 12 years previously because of cyanosis with a diagnosis of truncus arteriosus, so the patient was left untreated. Physical examination on recent admission revealed central cyanosis, clubbing of fingers and toes, a loud P2 on cardiac auscultation in association with mid-systolic and holo-diastolic murmurs over the chest. CXR showed cardiomegaly, increased pulmonary blood flow and pulmonary hypertension (Fig. 1).

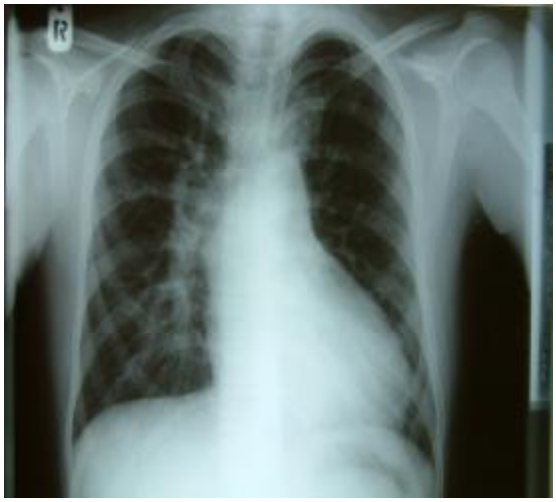

Fig. 1. CXR revealed cardiomegaly, increased pulmonary blood flow and pulmonary hypertension.

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Transthoracic echocardiography revealed double inlet single ventricle with LV morphology (Fig. 2) associated with levo-malposition of the great vessels.

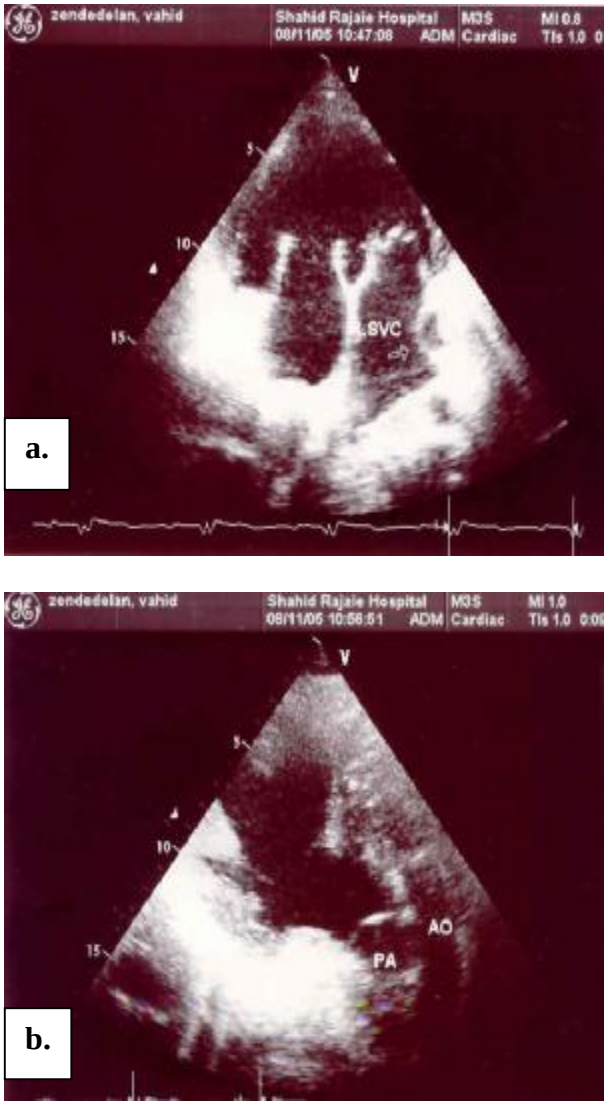


Fig. 2. Apical echo views showed double inlet single LV with dilated coronary sinus suggestive of persistent LSVC (a) and levo-malposition of great arteries (b).

There was an aneurysmally dilated great vessel which was located centrally and defined as pulmonary artery with moderate to severe pulmonary insufficiency (Fig. 3) and severe pulmonary hypertension (mean PAP = 85 mmHg).

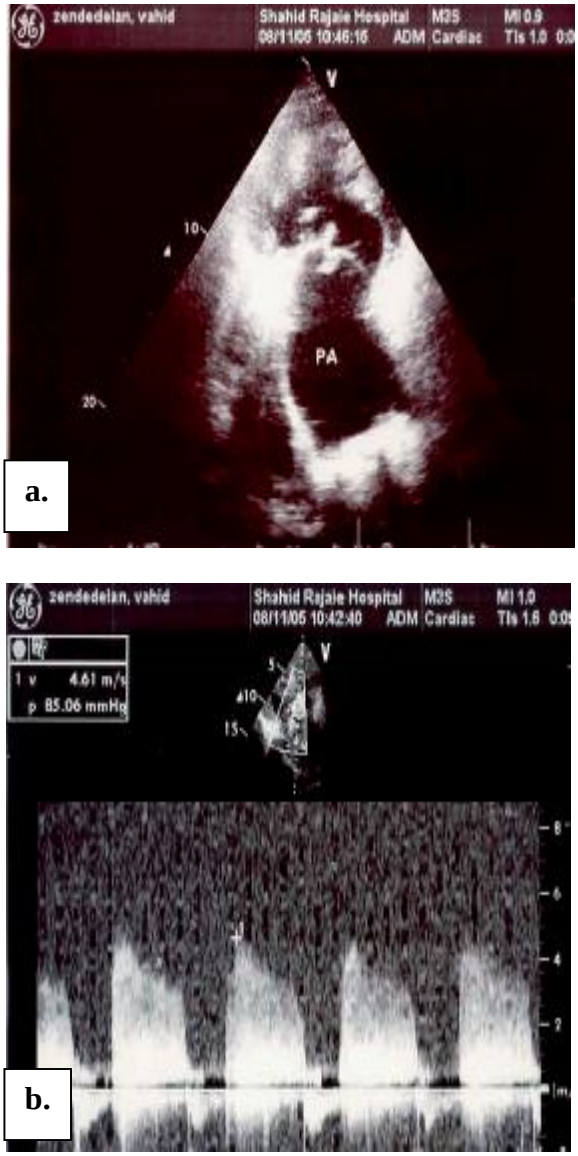
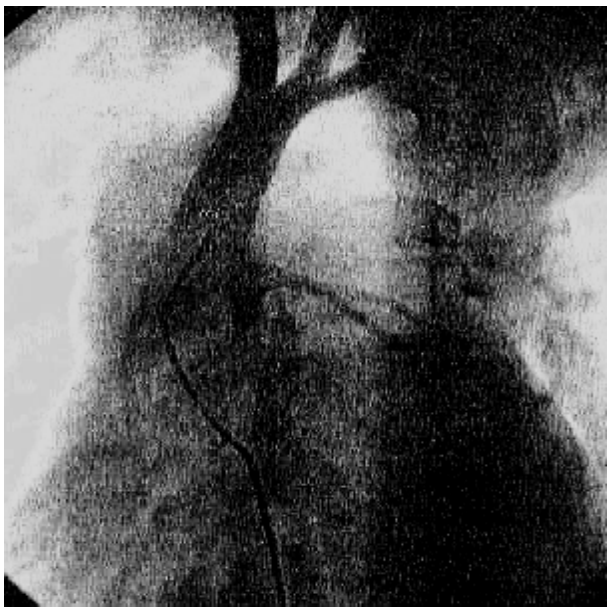


Fig. 3. 2D echocardiography revealed aneurysmal dilation of centrally located pulmonary artery (a), CW Doppler showed significant pulmonary hypertension (b).

The aorta was small sized and anteriorly located. The patient underwent repeated cardiac catheterization and all the above-mentioned data were confirmed. In addition, interrupted aortic arch (type A) was diagnosed; the descending aorta was supplied by the pulmonary artery via a giant PDA (Fig. 4).

a.



b.

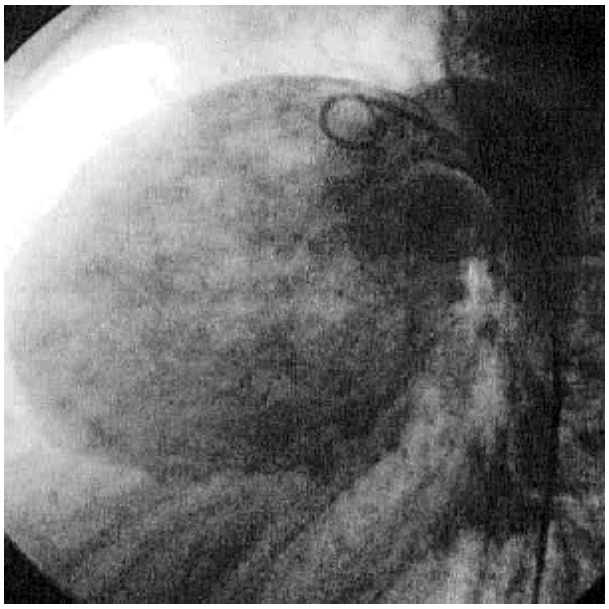


Fig. 4. Aortography (a) showed interruption of aortic arch after left subclavian artery take off (type A) and descending aorta is supplied by aneurysmally dilated pulmonary artery via a giant PDA (b).

Discussion

Interrupted aortic arch (IAA) is a rare congenital malformation of the aortic arch, which occurs in 3 per million live births.[1]

This anomaly is defined as a loss of luminal continuity between the ascending and descending portions of the aorta[2] which entails a very poor prognosis without surgical treatment. In most cases, IAA is associated with an intracardiac malformation such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve, left ventricular outflow tract obstruction, or aortopulmonary window. Interrupted aortic arch was initially described in 1778 by Steidele.[3] The first classification system introduced by Celoria and Patton[4] in 1959 is still used almost universally. This system describes and classifies the site of the aortic arch discontinuity, which may be distal to the left subclavian artery (type A); between the left carotid and left subclavian arteries (type B); or between the innominate and left carotid arteries (type C). The most common type is B (53%), followed by A (43%) and C (4%). In infants, its clinical presentation involves severe congestive heart failure; if the condition is left untreated, 90% of the affected infants die at a median age of 4 days.[11] In the few documented cases in adults, the presentation ranges from a lack of symptoms to limb swelling with differential blood pressures in all extremities. Substantial collateral circulation must be present to maintain flow and enable survival. However, collateral vessels are subject to atrophy and atherosclerosis, which can lead to other challenging problems.[12]

In adults, IAA is so rare that our review of the literature revealed only 12 cases. Nine of the patients underwent surgical repair.[1,5–10]

IAA is mostly diagnosed in early childhood due to congestive heart failure and differential cyanosis, but in our patient late diagnosis was secondary to double inlet LV with resultant mixing of saturated and unsaturated blood at the ventricular level and generalized cyanosis rather than differential cyanosis. Another cause of misdiagnosis was the very large central pulmonary artery, which mimicked truncus arteriosus.

The patient was not a candidate for any surgical intervention, and heart and lung transplantation was recommended.

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