

Aortic atresia with normal sized left ventricle

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Abstract

Aortic atresia with an associated ventricular septal defect and adequate sized left ventricle is extremely rare. We present two cases in which an alternate diagnosis was suggested on echocardiography because the hypoplastic aortic trunk was missed due to its small caliber. The final diagnosis was, however, clinched on dual source computed tomography, which not only showed the thin aortic trunk but also clearly depicted the coronary artery origins from the hypoplastic aortic root. To the best of our knowledge, use of multi-detector computed tomography in aortic atresia with well developed left ventricle has not been reported in literature till date.

Key words: Aortic atresia; left ventricle; multi-detector computed tomography; ventricular septal defect

Introduction

Aortic atresia is usually associated with hypoplastic left ventricle (LV) and or mitral valve in majority of the cases. Approximately 4–7% of the cases may have an associated ventricular septal defect (VSD) with a reasonably developed LV and mitral valve due to the VSD providing an outflow for the LV.^[1] Multi-detector computed tomography (MDCT) evaluation acts as a problem solving tool in the diagnostic work up of aortic atresia.

Case Report

Here, we describe two cases of aortic atresia in infants, which were diagnosed after they underwent CT angiography on a dual source 64 slice CT scanner (Siemens, Germany).

The first case was of a 5-month old male child who was incidentally found to have parasternal systolic cardiac murmur

during his pre-surgical check-up for congenital cataract surgery. Antenatal ultrasound was unremarkable. He had minimal cyanosis with regular pulse. His chest and abdominal examination were unremarkable. Echocardiography revealed both ventricles to be normal sized with single arterial trunk from the right ventricle, which was seen filling both the pulmonary arteries and the descending thoracic aorta, with a diagnosis suggestive of truncus arteriosus.

The second case was of a 6-month old male child with failure to thrive, recurrent chest infections, and difficulty in suckling. Antenatal ultrasound was unremarkable. He had minimal cyanosis with a systolic murmur in left parasternal area. Abdomen and chest examination were normal. Echocardiography revealed smaller LV with multiple ventricular septal defects with a suspicion of truncus arteriosus. Separate semilunar valves were not identified on echocardiography in both the cases, however, it showed normal atrioventricular valves.

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Both cases underwent gated CT angiography on a dual source 64 slice CT scanner using intravenous nonionic contrast under mild sedation without any complications.

In the first case, CT angiography [Figure 1] showed aortic atresia with hypoplastic aortic root and ascending aorta of 3 mm diameter. The arch, arch vessels, and descending thoracic aorta were normal. Normal origin of coronary arteries was seen from the hypoplastic aortic sinuses. Incidentally, aberrant right subclavian artery from proximal descending thoracic aorta with retro-esophageal course was also seen. There was a large membranous VSD with good sized LV. The second case [Figure 2 and Video S1] also showed aortic atresia with almost similar appearance of hypoplastic aortic root and ascending aorta of 3 mm diameter with normal arch, arch vessels, and descending thoracic aorta. Coronary artery origins were normal from the small sinuses. This patient had a large mid muscular VSD with a fairly developed LV. Both the patients were referred to the cardiothoracic surgeon for a possible biventricular repair.

Discussion

Blood flow to the embryonic systemic and pulmonary circulation acts as a stimulus in the normal development

process.^[2] When normal flow is compromised by an atretic or stenotic aortic valve, the ascending aorta and the coronaries receive usually low-pressure diastolic retrograde flow via the ductus arteriosus and aortic arch. This results in hypoplastic aortic root and ascending aorta. Usually, blood flow through the arch and the descending aorta remains adequate which induces their development to normal diameter.^[3] The presence of associated VSD provides an effective outflow, thus, inducing adequate growth of otherwise hypoplastic LV and mitral apparatus.^[1]

The diagnosis of the aortic atresia has been made on echocardiography in almost all the cases reported in literature.^[4] MDCT has emerged as a good alternative noninvasive modality for evaluation of cardiac structures with high temporal resolution.^[5,6] It allows fast acquisition with minimal breathing artifacts. In addition, the cardiac motion is minimized with the gated acquisition. In our cases, the hypoplastic aortic trunk was missed on echocardiography due to its small caliber and a diagnosis of truncus was suggested. MDCT not only showed the thread-like aortic trunk but also clearly depicted the coronary artery origins from the hypoplastic aortic sinuses. To the best of our knowledge, MDCT in aortic atresia with reasonably developed LV has never been reported in literature.



Figure 1: Computed tomography angiography images (A-D) from the first case demonstrating hypoplastic ascending aorta (arrow in A and C) with normal descending thoracic aorta (asterisk in A and B). Note is made of large patent ductus arteriosus (arrow in B), aberrant right subclavian artery (asterisk in C), and large ventricular septal defect (asterisk in D). Normal coronary artery origin from the hypoplastic sinuses and their course (arrowheads in C and D) is well seen with good sized left ventricle. R, right common carotid artery; L, left common carotid artery; S, left subclavian artery; RC, right coronary; CX, Circumflex; LV, left ventricle

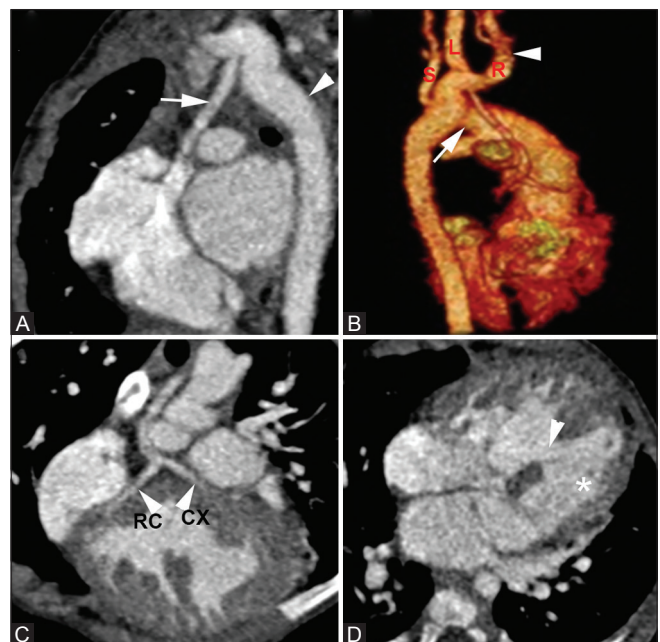


Figure 2 (A-D): Computed tomography angiography images (A-D) from the second case showing hypoplastic ascending aorta (arrow in A) with large patent ductus arteriosus (arrow in B), normal arch vessels (arrowhead in B), and descending thoracic aorta (arrowhead in A). Coronary arteries (arrowheads in C) are seen arising from the hypoplastic sinuses with mid-muscular ventricular septal defect (arrowhead in D) and fairly developed left ventricle (asterisk in D). R, right innominate artery; L, left common carotid artery; S, left subclavian artery; RC, right coronary; CX, Circumflex

Early surgical intervention is advocated in such cases to prevent development of intractable congestive heart failure and pulmonary vascular obstructive disease. Prognosis of aortic atresia associated with hypoplastic LV and mitral valve is generally very poor. However, this small subset of patients with VSD and adequate LV shows excellent long-term survival by primary biventricular repair.^[7]

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Conflicts of interest

There are no conflicts of interest.

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