■ *Case Reports* ■

Interruption of Aortic Arch in a Neonate: Diagnosis by Multi-slice Spiral Computed Tomographic Angiography

ZB **Hou,** X **Li,** CX **Wang**

Abstract   Interruption of the aortic arch is an uncommon congenital malformation commonly associated with other cardiovascular anomalies .We report a patient with type B interruption of the aortic arch diagnosed with multi-slice spiral computed tomographic angiography.

Key words   Angiography; Aortic arch; Computed tomography; Interruption

Case Report

A full term baby delivered by caesarean section because of oligohydramnios, was transferred to the neonatal unit of our hospital on day 5 of life with tachypnoea and cyanosis after crying. Physical examination revealed a grade 3 systolic murmur at the third and fourth left intercostal space and the femoral pulses were weak. The neonate had tachypnoea and was cyanosed. The blood pressure of his right upper limb was higher than that of the left. The temperature of the lower extremities was lower than the upper and both femoral pulses were weak. There was no history of heart disease in the family. The chest X-Ray showed patchy opacities in both upper lobes consistent with bronchopneumonia. Transthoracic echocardiography and Doppler study demonstrated the presence of ventricular septal defect (VSD) and atrial septal defect (ASD),

decreased flow velocity in the descending aorta, patent ductus arteriosus (PDA) with right to left shunt from the main pulmonary artery (MPA) to the descending aorta (DA) in systolic phase and in the opposite direction in the diastolic phase. Interruption of aortic arch (IAA) was suspected but could not be confirmed (Figure 1). The purpose of contrast-enhanced CT angiography was for further confirmation of the presence of interruption of aortic arch. The patient was sedated with oral chloral hydrate for the 16-slice spiral computed tomographic angiography (SIMENS SENSATION [120 Kv,150 mAs]). The contrast agent used was Lopamiro and injected at a speed of 1.5 ml per second. The 3-dimensional (3-D) reconstruction images combined with echocardiography revealed sub-pulmonary trunk ventricular septal defect (SPVSD) with a maximum diameter about 0.7 cm, an ASD located in the area of the fossa ovalis with a maximum diameter of about 0.6 cm, dilated pulmonary artery (PA) (Figures 2A, C & D) and interrupted aortic arch between the left common carotid artery and the left subclavian artery consistent with type B interrupted aortic arch (Figure 2C). The descending aorta (Figure 2B) was connected with the pulmonary artery through the PDA (Figure 2A). Surgical treatment was planned but the family could not afford the medical expenses. The parents opted for conservative treatment and the patient died of acute heart failure at around 6 months of age.

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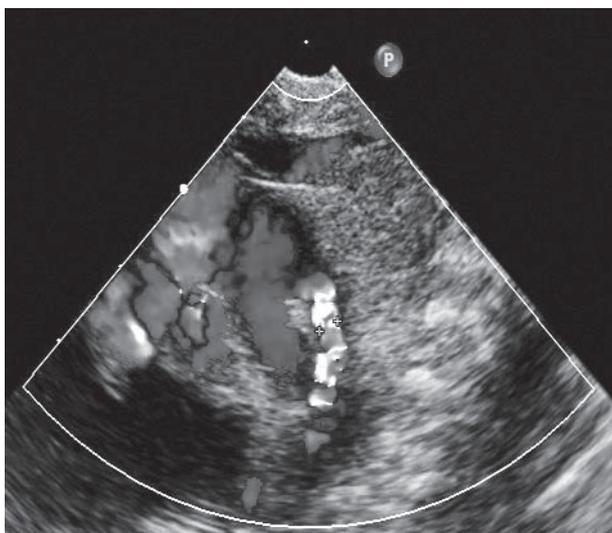
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Figure 1   The echocardiography showed the blood direction was from the descending aorta to the main pulmonary artery through the patent ductus arteriosus in diastolic,the succession from ascending aorta to descending aorta displayed not well.
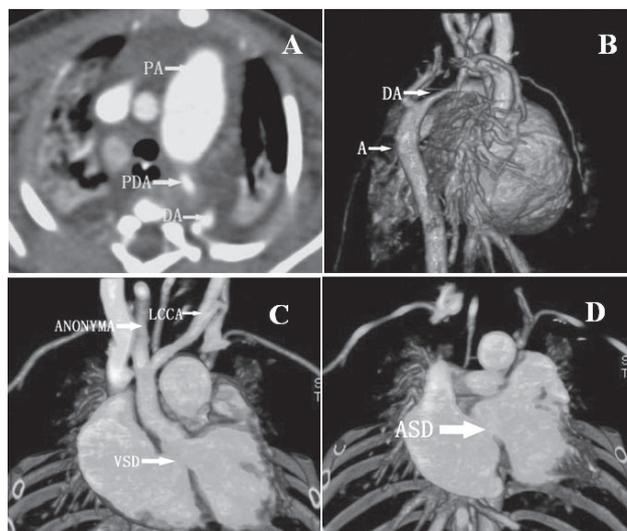

Figure 2   Dilation of pulmonary artery (PA), descending aorta (DA) was connected with pulmonary artery through descending aorta (DA)" (Figures A & B). Aortic arch disappeared, ascending aorta was divided into anonyma and left common carotid artery (LCCA), ventricular septal defect (VSD), atrial septal defect (ASD) (Figures C & D).

Discussion

In the fully developed human with a normal left aortic arch, the derivatives of the primitive left and right fourth arches form the distal aortic arch (the segment between the left common carotid artery and left subclavian artery). In the sixth to seventh week of the embryonic period, the aortic arch is derived from the left dorsal aorta or the fourth arch. The blood supply to the upper half of the body depends on left ventricular outflow and that to the lower half of the body depends on the right ventricular output through the patent ductus arteriosus. Therefore, the clinical manifestation is a difference in the pulse volume between left and right limbs or upper and lower limbs.

IAA is defined as the discontinuity of the aortic arch between the ascending and descending aorta and is a rare conotruncal congenital cardiovascular malformation and constitutes one of the most rapidly lethal forms of congenital cardiovascular disease.[1-3] It occurs in 1:10,000 births, which could be associated with several chromosomal abnormalities or as a result of a monogenic defect.[4] Isolated IAA is rare and the condition it usually associated with other cardiovascular malformations and accounts for about 1% of the patients with congenital heart defects.[5] The site

of interruption of the aortic arch has been used for the classification.[6] IAA is divided into 3 pathological types: the interruption classified as type A is located on the distal end of left subclavian artery; type B interruption is between left common carotid artery and left subclavian artery and in type C, the interruption is between the innominate and left common carotid arteries. According to published literature,[7] type A is the most common and type C is rare in Chinese. Patients with IAA type B can only survive with an obligatory PDA and patients frequently also have an associated VSD and a malaligned ventricular septum with potential for subaortic stenosis. In type B, interruption which is the condition found in the present reported case, the left fourth arch is always abnormal because interruption of the arch occurs at this location, and the right fourth arch is also frequently abnormal. Similarly, in cases of interruption of the right sided arch, the fourth arches are often involuted bilaterally, with type B interruption and an aberrant or isolated left subclavian artery also being found.[8]

Echocardiogram is the method of first choice to detect IAA. Suprasternal echocardiographic view can demonstrate a loss of luminal continuity between the ascending and descending aorta, absence of bloodflow in the distal end of aortic arch, and other associated malformations.

Echocardiography has been shown to be superior to computed tomography in demonstrating intracardiac malfomation and bloodflow in the great vessels. However, echocardiography is influenced by the technical level of the operator, ultrasonic near-field, and interference by chest wall and air in the lungs. It can be difficult at times to clearly demonstrate the presence or absence of continuity between the aortic arch and descending aorta. Although digital subtraction angiography (DSA) is the gold standard for detecting angiocardiopathy, it is invasive and it may not be the imaging modality of choice in sick neonates with aortic interruption who are usually already in cardiac and renal failure. DSA is also more expensive than CT and other noninvasive techniques.

Multi-slice spiral computed tomographic angiography (MSCTA) offers a short scanning time, high spatial resolution and adequate temporal resolution. This technique allows the retrospective reconstruction at any phase of a cardiac cycle and is thus suitable for the detailed evaluation of cardiac anatomy. Three-dimensional reconstruction can clearly delineate the spatial anatomical relationships of the large blood vessels. Furthermore, MSCTA can display real-time 3D anatomic structure. Fast 3D reconstruction and real-time display are helpful in improving the planning for surgical management. We propose that MSCTA is comparable to DSA for the diagnosis of certain cardiovascular malformations in infancy if properly used. However, as compared to echocardiography, MSCTA has limitations in displaying intra-cardiac malformations and changes in haemodynamics. In addition, because of the fast heart rate in neonates, the coronary arteries of heart are not well displayed. MSCTA will become an alternative method to X-ray angiography and other noninvasive techniques and will have an important role in the diagnosis of cardiovascular malformations in young infants.

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