Case Report

4D-flow MRI of double aortic arch in a 14-year-old patient

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Abstract: Double aortic arch is a rare congenital anomaly. It is usually diagnosed and surgically corrected at an early age due to symptoms as dyspnea and dysphagia caused by an obstruction of trachea and/or esophagus in the vascular ring. We present the case of an asymptomatic 14-year-old patient with complete double aortic arch as demonstrated by CMR. Blood flow in the right and left aortic arch was visualized and quantified by 4D-flow MRI.

Keywords: Double aortic arch (DAA); cardiovascular magnetic resonance (CMR); 4D-flow MRI

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A 14-year-old girl with suspected aortic anomaly was referred to the outpatient department. She reported no impairment of physical activity, or incidents of dysphagia or dyspnea.

Transthoracic echocardiography showed double aortic arch (DAA) with suspected incomplete left arch.

Cardiovascular magnetic resonance (CMR) imaging revealed complete DAA (*Figure 1* and *Video 1*), forming a non-interrupted vascular ring with carotid and subclavian arteries separately arising from each arch (*Figures 1,2* and *Video 2*). Trachea and esophagus were not compressed by the vascular ring. Furthermore, 4D-flow MRI (*Figure 3*) proved blood flow in both aortic arches. A flow distribution of blood from the ascending aorta [effective stroke

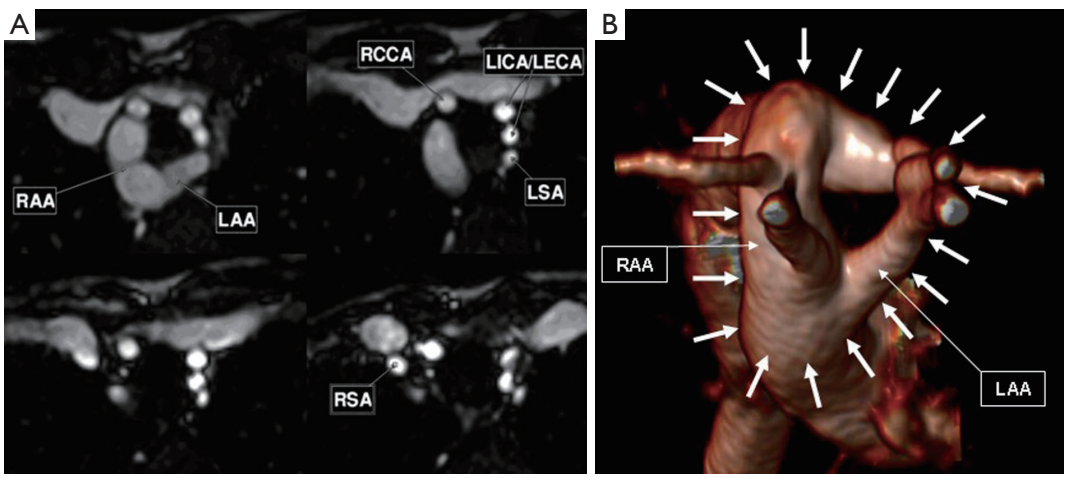

Figure 1 Double aortic arch with carotid and subclavian arteries, cranial view. (A) Transversal steady-state free-precession images; (B) Three-dimensionally reconstructed magnetic resonance angiogram. LAA, left aortic arch; LECA, left external carotid artery; LICA, left internal carotid artery; LSA, left subclavian artery; RAA, right aortic arch; RCCA, right common carotid artery; RSA, right subclavian artery.

Video 1 Three-dimensionally reconstructed magnetic resonance angiogram of aorta-nod.

Video 2 Three-dimensionally reconstructed magnetic resonance angiogram of aorta-spin.

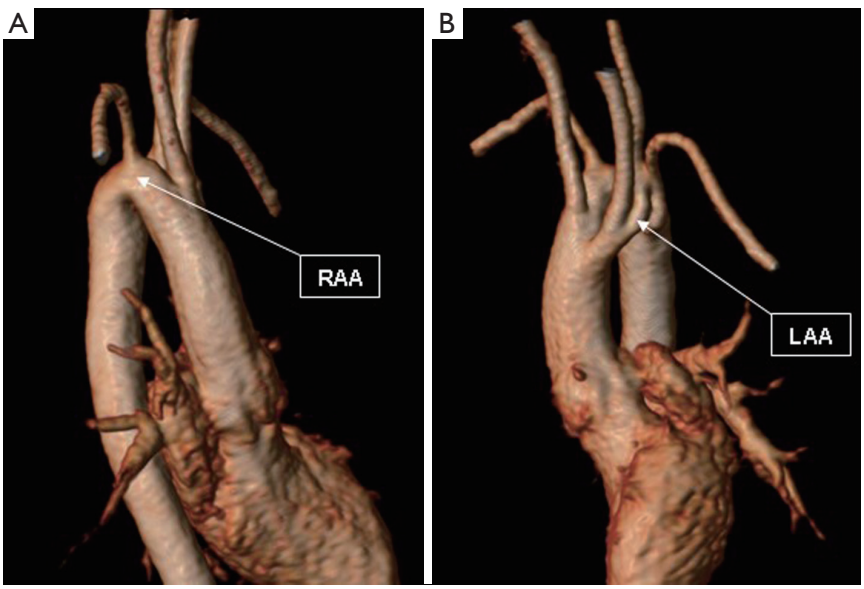

Figure 2 Double aortic arch with carotid and subclavian arteries. (A) Three-dimensionally reconstructed magnetic resonance angiogram, right anterior view showing right common carotid artery and right subclavian artery arising from the larger right aortic arch; (B) Three-dimensionally reconstructed magnetic resonance angiogram, left anterior view showing left subclavian artery, and left intern and extern carotid arteries separately branching off the smaller left aortic arch. LAA, left aortic arch; RAA, right aortic arch.

volume (SV eff.) 48 mL, peak velocity (Vmax) 1.1 m/s] of approximately 4/5 streaming through the larger right (SV eff. 38 mL, Vmax 0.9 m/s), and 1/5 through the smaller left aortic arch (SV eff. 9 mL, Vmax 0.6 m/s) was demonstrated.

DAA is a rare congenital anomaly which can appear in several different anatomical variations (1). It is usually diagnosed and surgically corrected at an early age due to symptoms caused by an obstruction of trachea and/or esophagus in the vascular ring. Single cases of elderly patients with late diagnosis of DAA have been described in the literature (2-8). In our case, CMR showed complete DAA with effective blood flow in both the right and the left aortic arch as visualized and quantified by 4D-flow MRI. Consistent with the absence of symptoms as dyspnea and

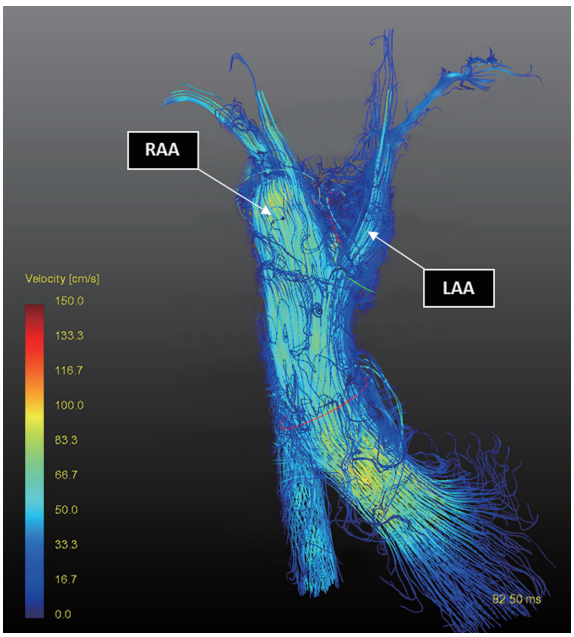

Figure 3 4D-flow image of double aortic arch, anterior view.

dysphagia, there were no signs of compression of trachea and esophagus in the CMR. Therefore, surgical repair was not indicated.

CMR is a valuable tool for diagnosis of cardiovascular malformations as DAA. Furthermore, 4D-flow MRI may enable visualization of blood flow within the vessels and provide important functional information considering flow distribution, velocities and flow patterns at different anatomical regions over time. As evaluated by Karmonik *et al.*, computational fluid dynamics (CFD) in addition to MRI might be useful for pretreatment planning in patients with aortic diseases (9).

In conclusion, we recommend CMR including 4D-flow MRI for diagnosis and clinical decision making in DAA. In patients with only minor or no symptoms attributable to DAA but planned cardiovascular surgery for other reasons, the so far unclear benefit of additional DAA repair may in the future be further investigated by simulating treatment scenarios through CFD based on MRI.

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References

1. Kellenberger CJ. Aortic arch malformations. Pediatr Radiol 2010;40:876-84.
2. Kypson AP, Anderson CA, Rodriguez E, et al. Double aortic arch in an adult undergoing coronary bypass surgery: a therapeutic dilemma? Eur J Cardiothorac Surg 2008;34:920-1.
3. Fernandez-Valls M, Arnaiz J, Lui D, et al. Double aortic arch presents with dysphagia as initial symptom. J Am Coll Cardiol 2012;60:1114.
4. Gami AS, Ammash NM. Images in cardiovascular medicine. Double aortic arch. Circulation 2004;109:2370-1.
5. Ikenouchi H, Tabei F, Itoh N, et al. Images in cardiovascular medicine. Silent double aortic arch found in an elderly man. Circulation 2006;114:e360-1.
6. Kothari SS, Roy A, Sharma G. Left ventricular pseudoaneurysm in a child. Heart 2005;91:773.
7. Koz C, Yokusoglu M, Uzun M, et al. Double aortic arch suspected upon transthoracic echocardiography and diagnosed upon computed tomography. Tex Heart Inst J 2008;35:80-1.
8. Yilmaz M, Tok M, Cengiz M. Asymptomatic balanced-type double aortic arch in an elderly patient: a case report. Heart Surg Forum 2007;10:E297-8.
9. Karmonik C, Partovi S, Davies MG, et al. Integration of the computational fluid dynamics technique with MRI in aortic dissections. Magn Reson Med 2013;69:1438-42.

