

[PICTURES IN CLINICAL MEDICINE]

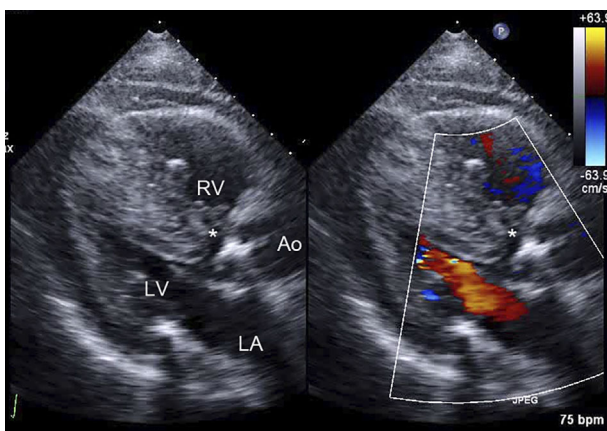
Right-sided Aortic Arch Associated with Congenital Heart Disease

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Key words: right-sided aortic arch, bicuspid aortic valve, ventricular septal defect

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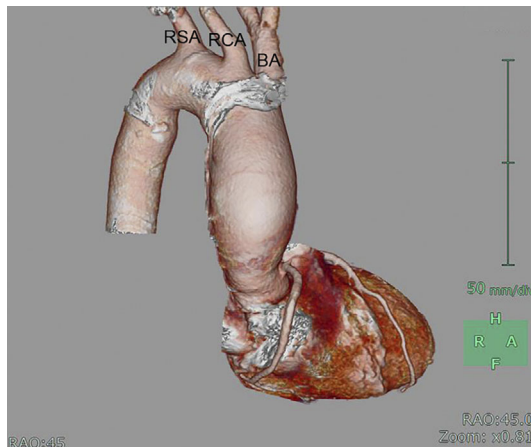
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Picture 1.



Picture 2.



Picture 3.



Picture 4.

A 68-year-old woman was transferred to our hospital to determine the cause of fainting. Echocardiography demonstrated severe aortic stenosis (AS, peak velocity, 5.3 m/s) caused by a bicuspid aortic valve along with a small pe-

rimembranous ventricular septal defect (VSD; Picture 1). Computed tomography (CT) confirmed a bicuspid valve (Picture 2: arrowhead) and revealed a right-sided aortic arch (RAA) with mirror-image branching of a normal circumflex

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aortic arch (Picture 3, 4: asterisk). In addition, CT revealed a thoracic ascending aortic aneurysm (TAA) caused by post-stenotic dilatation for severe AS (Picture 3, 4: arrow). The patient underwent surgical replacement for AS and repair for VSD and TAA. Since the surgery, she has not fainted. RAA is a rare congenital defect of the aorta, and the complications associated with AS and VSD are unusual (1). Type I RAA is typically associated with tetralogy of Fallot and truncus arteriosus (2). The cause of RAA is largely unknown. A deletion in chromosome 22q11 is associated with a 24% incidence of isolated anomalous branching of the aortic arch (3). However, our patient did not have this deletion. Further studies are warranted to determine whether these diseases are incidental or involve the maldevelopment of cardiac genes, such as *TBX1*.

The authors state that they have no Conflict of Interest (COI).

References

1. Cina CS, Althani H, Pasenau J, Abouzahr L. Kommerell's diverticulum and right-sided aortic arch: a cohort study and review of the literature. *Journal of vascular surgery* **39**: 131-139, 2004.
2. Arazinska A, Polgaj M, Szymczyk K, Kaczmarek M, Trebinski L, Stefanczyk L. Right aortic arch analysis - anatomical variant or serious vascular defect? *BMC Cardiovascular Disord* **17**: 102, 2017.
3. McElhinney DB, Clark BJ 3rd, Weinberg PM, et al. Association of chromosome 22q11 deletion with isolated anomalies of aortic arch laterality and branching. *J Am Coll Cardiol* **37**: 2114-2119, 2001.

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