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IMAGES in PAEDIATRIC CARDIOLOGY

Muratori D[1], Meani P[2], Quattrocchi G[1], Pedrotti P[1]. Association of unicuspid unicommissural aortic valve and complex congenital heart disease depicted by cardiac magnetic resonance.. Images Paediatr Cardiol 2016;16(3):5-8.
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Patient

A 12-year-old male child was referred for follow-up cardiac magnetic resonance (CMR) of complex congenital heart disease, characterized by aortic decoarctation, interventricular and interatrial septal defects (VSD and ASD) closure and bicuspid aortic valve.

CMR showed unicuspid unicommissural aortic valve with normal valve area and without significant regurgitation. A sub-aortic spur was detected, without outflow obstruction. There were no signs of recoarctation nor residual septal defects; both ventricles were of normal size and function (figures 1-3).

Figure 1. Aortic valve, CMR cine image, systolic still frame, showing unicuspid unicommissural aortic valve.

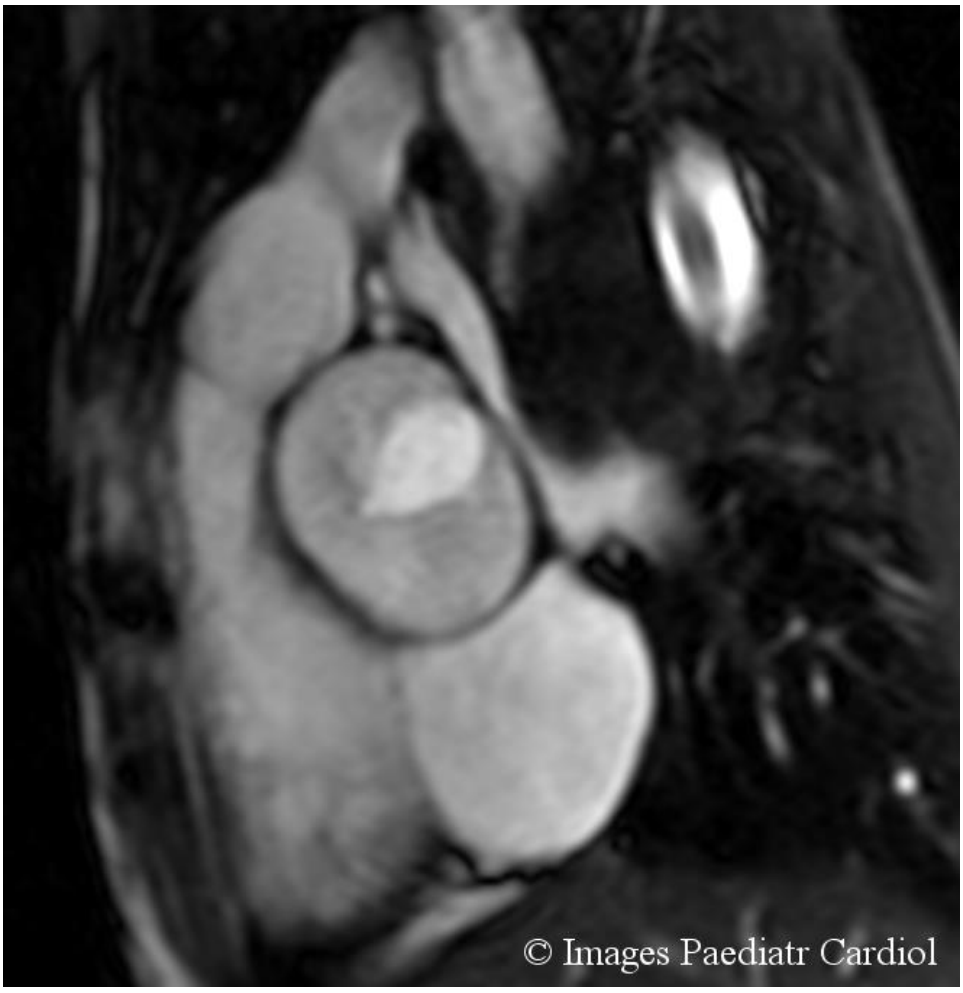


Figure 2. Left ventricular outflow tract, CMR cine image, systolic still frame, showing eccentric aortic opening (short arrow) and subaortic spur (long arrow).

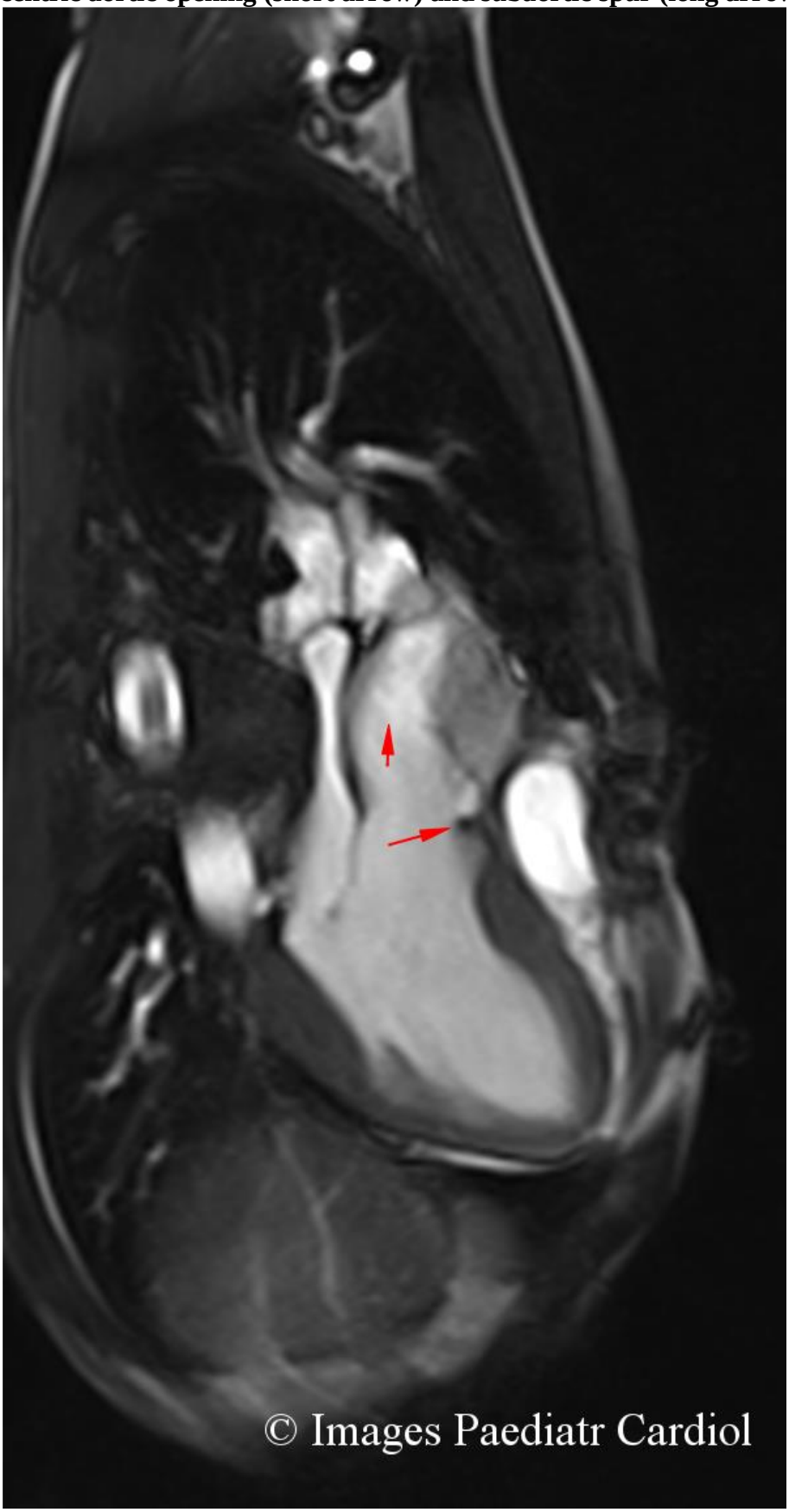
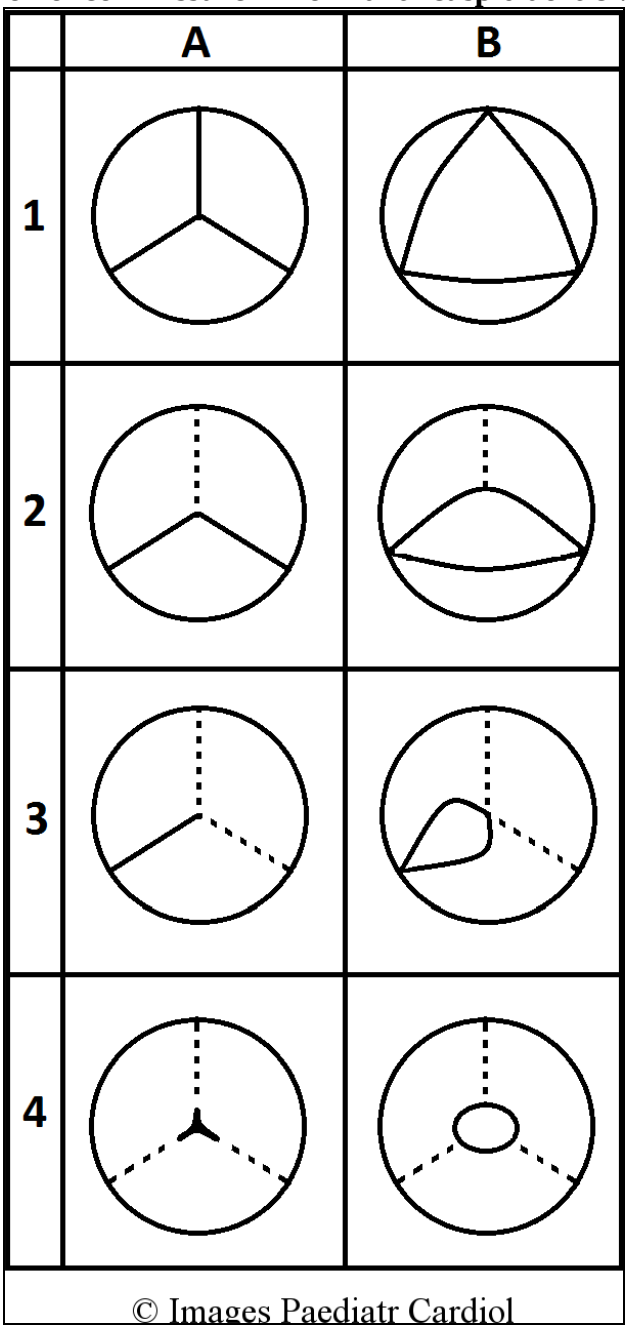


Figure 3. Schematic illustration of the anatomic variations of aortic valve. (A) closed valve; (B) open valve. 1) Normal tricuspid aortic valve. 2) Bicuspid aortic valve. 3) Unicuspid unicommissural aortic valve. 4) Unicuspid acommissural aortic valve. Dotted lines indicate the location of commissure in normal tricuspid aortic valve.



Discussion

Unicuspid aortic valve (UAV) is a rare congenital anomaly with an incidence of 0.02% in the adult population as estimated by echocardiography.[1] It is usually associated with aortic stenosis or regurgitation.

Two forms of UAV have been described: unicuspid unicommissural aortic valve, in case of presence of a lateral attachment to the aorta and unicuspid acommissural aortic valve, in case of absence of this attachment.[2]

Transthoracic and transesophageal echocardiography (TTE and TEE) are the gold standard imaging diagnostic techniques for the assessment of aortic valve abnormalities.[3] However, in a recent

Canadian study on 42 patients who underwent aortic valve surgery for stenosis or regurgitation, UAV had been identified by pre-operative TEE only in 69% of the cases.[4]

CMR has become pivotal in the clinical evaluation of aortic diseases, especially in the context of congenital heart disease.[3] Only a few cases of UAV with coexisting cardiac anomalies detected by echocardiography and CMR have been reported.[1-5]

In our patient, pre-operative echocardiographic assessment had not identified UAV. Moreover, to the best of our knowledge, the combination of UAV, aortic coarctation, and VSD and ASD has not been previously described. CMR allowed the identification of UAV, together with follow up evaluation of complex congenital heart disease, confirming the importance of this diagnostic tool in this clinical setting.

References

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