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IMAGE

An Unusual Finding of a Double Orifice Mitral Valve in a Patient With Holt-Oram Syndrome

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A 41-year-old lady presented to the Emergency Department with recurrent, disabling paroxysms of supraventricular tachycardia alternating with third degree atrioventricular block. Her history was notable for Holt-Oram Syndrome with previous childhood atrial septal and ventricular septal defect repair, and she had been lost to follow-up after her surgery. Transthoracic echocardiography revealed an incidental finding of an abnormal mitral valve with two distinct equally sized orifices separated by a fibrous bridge consistent with a double orifice mitral valve (DOMV) (Figures 1 and 2). The valve was well functioning with no significant mitral stenosis or incompetence; continuous wave Doppler revealed a short pressure half-time (<50 milliseconds through each orifice), with no significant regurgitation on colour Doppler interrogation, and pulse-wave Doppler at the leaflet tips of both orifices demonstrated similar antegrade peak velocities (E velocity ~80 cm/sec). The patient underwent an electrophysiologic study which demonstrated a focal

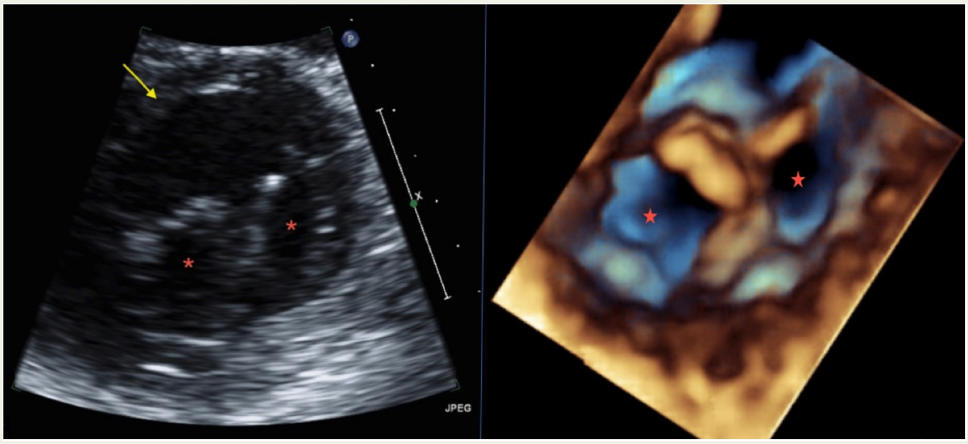

Figure 1 Parasternal short axis view of the left ventricle showing 2D (left) and 3D (right) views. Yellow arrow = interventricular septum; red stars = mitral valve orifices (2).

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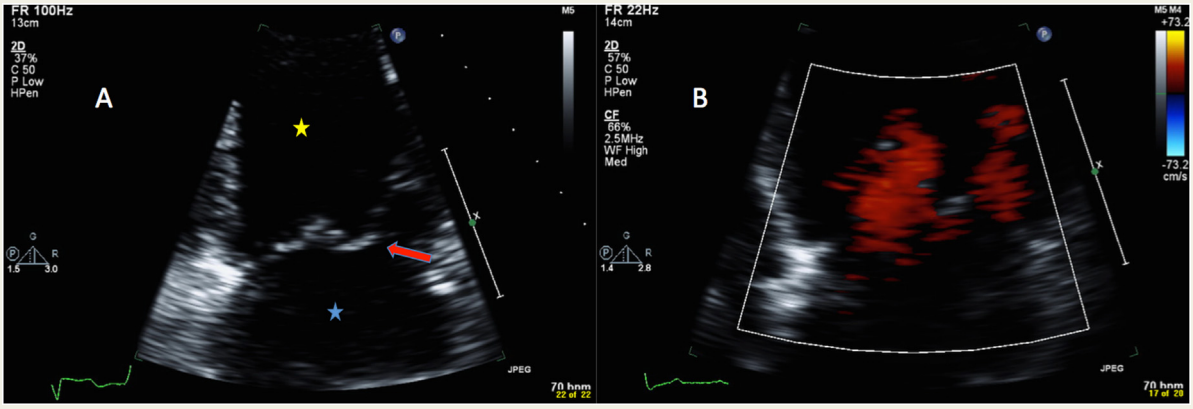

Figure 2 Apical 4-chamber view. **A** shows the left atrium (blue star), left ventricle (yellow star) and mitral valve (red arrow). In **B**, colour Doppler interrogation of the mitral valve during diastole demonstrates two parallel inflow jets.

tachycardia originating at the inferolateral mitral annulus that was successfully ablated, and post-ablation has remained symptom-free.

Holt-Oram Syndrome (HOS) is a rare autosomal dominant disorder that causes abnormalities of the upper limbs and heart, in particular conduction abnormalities and structural defects. DOMV is a rare cardiac congenital anomaly often diagnosed incidentally during investigation for other congenital heart disease. There have been reports describing the presence of DOMV and atrial arrhythmias, however the combination of DOMV and HOS has not been described. Furthermore, we have demonstrated a localised annular site of tachycardia which may represent a link in the pathophysiology of congenital mitral valve disease and arrhythmias.

Given the possibilities of mitral incompetence or stenosis with a structurally abnormal valve, patients with DOMV require regular echocardiographic surveillance. Patients with DOMV and palpitations should be considered for further investigation and possibly invasive assessment.

To our knowledge this is the first case of DOMV coinciding with HOS, and given the propensity of HOS to cause cardiac anomalies the two conditions are possibly associated in this patient.

Contributions

1. Dr. Kunal Pradip Verma: design of work, acquisition of images, drafting of manuscript.
2. Dr. Stewart Healy: final approval of the version to be published.
3. Dr. Justin Teng: review of manuscript, acquisition of images.
4. Dr Nitesh Nerlekar: design of work, final approval of the version to be published.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at https://doi.org/10.1016/j.hlc.2018.11.017.