AN ELUSIVE QUEST TO EXPLAIN THE WORSENING “PULMONARY HYPERTENSION” IN A CASE OF KNOWN STABLE RESTRICTIVE VENTRICULAR SEPTAL DEFECT

Poster Contributions
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Gerbode ventricular septal defect (VSD) is a rare communication between the left ventricle and right atrium. It is generally congenital in origin but on rare occasions is an acquired defect. Gerbode defect can either arise from a direct communication between LV and RA with an atrio-ventricular perimembranous VSD or an indirect communication via defect in the septal leaflet of the tricuspid valve (TV) in presence of a perimembranous VSD.

A 39yo female with a known history of congenital small restrictive VSD and worsening right ventricular systolic pressure (RVSP) since history of prolonged infection in 2008 was being evaluated for her unexplained pulmonary hypertension. She presented to our institution with fevers and was found to have Streptococcal bacterial endocarditis. A TEE demonstrated vegetation on the tricuspid valve with moderate to severe eccentric tricuspid regurgitation (TR), mild RV dilatation and a small restrictive VSD. She responded to antibiotics and remained hemodynamically stable. However, the reason behind her worsening TR, elevated RVSP and RV dysfunction remained unexplained despite workup for pulmonary hypertension.

Since her hemodynamics and RV function had worsened significantly from her TTE ~6 months earlier, cardiac MRI (CMR) was undertaken to further evaluate the nature of her valvular pathology as echocardiograms failed to reveal a clear explanation for worsening RVSP. CMR demonstrated a TV septal leaflet perforation with severe eccentric TR jet. The membranous portion of the interventricular septum was aneurysmal with an abnormal communication between the subaortic region of the LVOT and the right atrium. The QP:QS was 1.4:1. CMR identified the Gerbode defect and TV septal perforation, which had not been identified with prior evaluations with echocardiogram. Her worsening hemodynamics (“PHTN”) was most likely due to acquired Gerbode defect post endocarditis. This case demonstrates the value of CMR in accurate evaluation of VSD in a patient with worsening RVSP. CMR clearly demonstrated the Gerbode defect, likely acquired, missed in her prior echocardiograms. Accurate identification of pathology prompted surgical management with VSD closure and TV repair.