PFO DEVICE CLOSURE IN A PATIENT WITH HYPOPLASTIC RIGHT VENTRICLE AND CYANOSIS

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History and physical:

A 23 year old lady with history of pulmonary balloon valvuloplasty at childhood presented with progressive exertional dyspnea and palpitation. Physical examination was remarkable for cyanosis, finger clubbing and oxygen saturation at room air of 70%.

Imaging:

Transthoracic echocardiography showed a small hypertrophied RV cavity. Tricuspid annulus measured about 2.8cm (lower limit of normal s) and right ventricular outflow tract was hypoplastic (RVOT: 1.06cm) with no stenosis. There was mild to moderate RV dysfunction and a large stretched PFO with bidirectional shunt

Cardiac magnetic resonance (CMR) demonstrated RV size of 44cc/m2 in favor of a small RV size or borderline RV hypoplasia with severe RV hypertrophy, hypoplastic main pulmonary artery and few small aorto-pulmonary collaterals.(Figure1)

Indication for intervention:

Regarding cyanosis and clinical symptoms, the patient was evaluated for possibility of device closure of interatrial communication.

Intervention:

Cardiac catheterization revealed RV pressure of 40/0-10 mmHg and pulmonary artery pressure of 30/12 mmHg. We decided to evaluate whether the patient required the interatrial communication to maintain adequate cardiac output. The PFO was occluded with a balloon catheter for twenty minutes during which close hemodynamic monitoring was performed. No drop in systemic blood pressure and no significant rise in right atrial pressure and RV end diastolic pressures were observed throughout the occlusion test. The systemic oxygen saturation also improved. Device closure of the PFO was done successfully. After the procedure the patient became clinically acyanotic with oxygen saturations between 90-92%.

In the follow up visit three weeks after the procedure the patient remained asymptomatic and transthoracic echocardiography showed that RV function had improved compared to the pre-procedural study. There was also no increase in tricuspid regurgitation severity. (Figure2)

Learning points of the procedure:

Isolated RV hypoplasia with ASD or PFO is a rare congenital heart disease. Trans-catheter closure of atrial septal defect in the presence of RV hypoplasia is challenging and needs special considerations to prevent RV failure, increase in caval venous pressures and worsening clinical symptoms after the procedure. The ability of the right ventricle to maintain adequate forward flow and proper adjustment to the venous return is critical. There is limited experience on the best management strategy in these patients. Few cases with isolated RV hypoplasia and ASD have been reported in the literature and temporary occlusion of the ASD/PFO with a balloon catheter and hemodynamic monitoring has been proposed to identify the suitable candidates for device closure of the inter-arterial defects. Patients who are intolerant of the device closure generally require surgical repair with concomitant cavopulmonary anastomosis including Glenn or Fontan type procedures.

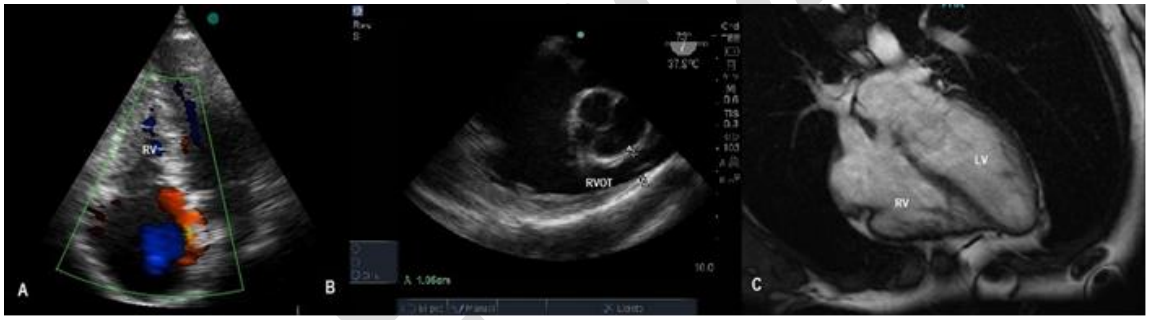

Figure 1: Transthoracic echocardiogram in four chamber view depicting hypoplastic right ventricle(A),Transesophageal echocardiogram showing underdeveloped RV infundibulum(B, arrow),Cardiac magnetic resonance showing small RV cavity(C)

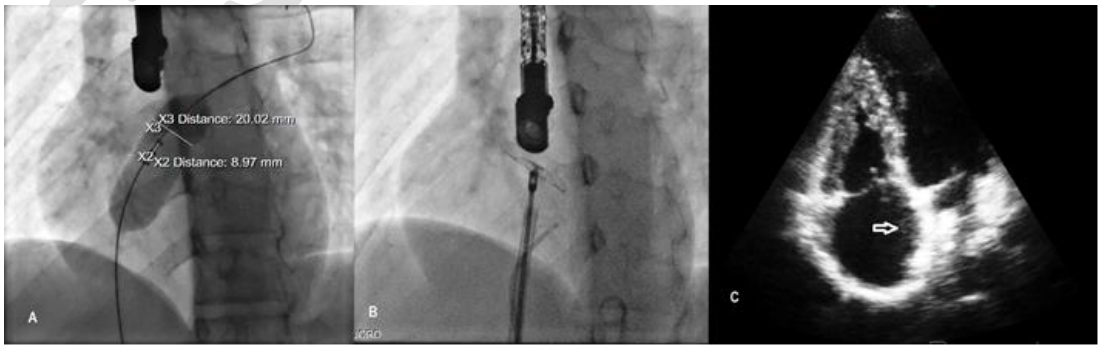

Figure 2: Balloon test and sizing balloon occlusion of the stretched PFO (A), Device closure of the defect (B), Post procedural echocardiogram with device in proper position(C, arrow)