AP Target Symposium

AP Target Symposium 2 (I-APT2)
Dealing with the borderline Right Ventricle - Fontan vs One-and-a-Half Ventricle Repair vs Biventricular Repair: what are the criteria and how to get there –

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Fri. Jul 7, 2017 4:15 PM - 5:45 PM ROOM 3 (Exhibition and Event Hall Room 3)

4:15 PM - 5:45 PM

[I-APT2-04]Decision-making process in theatre: Ebstein’ s anomaly (One and a half repair in Ebstein’ s anomaly)

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Since the original contribution of Billingsley et al[1] about the one and a half repair in hypoplastic right ventricle, the indications of this strategy have been extended to a variety of congenital heart disease. The clinical situation for the one and a half repair include right ventricular outflow obstruction with a hypoplastic right ventricle(RV), pulmonary atresia with intact ventricular septum, Ebstein’ s anomaly, complex atrioventricular septal defect, inlet VSD with straddling of tricuspid valve, and the complex anomaly with difficulty factors during intracardiac repair[2].
In patients with Ebstein’ s anomaly, additional bidirectional Glenn (one and a half reapir) with tricuspid valve repair or replacement achieves: it decrease the volume loading on the enlarged and dysfunctional RV, provides adequate preload to LV, reduces the hemodynamic stress on a complex TV repair, permits a more aggressive valve repair while avoiding valve stenosis. tolerates longer intervals between repeat TV operations for progressive TR and failing TV prostheses, and it provides less mortality[3-5].
Indication of one and a half repair in Ebsein’ s anomaly include severe cyanosis at rest, severe RV dilatation or dysfunction (cardiothoracic ratio >65%, RVEDV >250 mL/m2, RVEF <0.25), small, squashed, D-shaped left ventricle(leftward shift of IVS), postrepair RAP:LAP >1.5:1, postrepair low cardiac output (persistent metabolic acidosis, low urine output, increasing creatinine, poor peripheral perfusion, and low mean arterial blood pressure <50 mm Hg), stenotic TV orifice after repair(mean gradient >8mmHg)[4-8].
The patient should have good LV function and good pulmonary conditions: LVEDP less than 15mmHg, good pulmonary artery pressure (less than 18 – 20 mmHg), and transpulmonary gradients less than 10 mmHg[7]. For the good hemodynamic after one and half repair, the right ventricular outflow tract obstruction must be avoided and the pulmonary valve is needed[9].
Contraindications [6] for one and a half repair are profound RV dilatation or dysfunction (RV ejection fraction [EF] <10%), left ventricular dilatation with dysfunction (LV EF <25%), , mean PA pressure >20 mm Hg, pulmonary arteriolar resistance >4 Woods units, LVEDP or LA pressure >12 mm Hg, and significant pulmonary artery hypoplasia.
Concerns after one and a half repair include possible development of venoveno collaterals due to high CPV, facial suffusion, pulmonary arteriovenous fistulae, central venous thrombosis, and difficulty access for right ventricle for electrophysiology study, ablation, and pacing lead insertion.
The one and a half ventricle repair seems to be a valid alternative to Fontan and biventricular repairs in

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selected patients with Ebstein’ s anomaly.

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