POTTS shunt as a palliation for patients with idiopathic PAH

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Introduction:
Idiopathic pulmonary artery hypertension(IAPH) can present at any age from infancy to adulthood. It is a progressive disease and may become fatal in many patients. Literature has shown that survival of Eisenmenger's syndrome is much better as compared with patients with IAPH. POTTs shunt i.e side to side anastomosis between left pulmonary artery and aorta has been proposed to convert children with IAPH into Eisenmenger's syndrome . It has been shown to improve functional class and prolonged survival in few reports.
We report clinical outcomes of first 8 patients with IAPH who underwent POTTs shunt at our institute in last two years
Materials and Methods:
This is a prospective study where 8 patients (age= 9 months-35 years) with idiopathic pulmonary artery hypertension who were on maximal medical therapy and in functional class IV were enrolled. Parents/patients were explained the nature of the disease, the procedure and the implications of the same. Clinical symptoms of syncope, functional class, signs and symptoms of right heart failure, upper and lower limb oxygen saturation was looked for in all the patients. Detailed echocardiography was performed in all the patients. Accurate measurements of pulmonary artery pressures and right ventricular size and function was assessed. PA systolic pressure (PASP), PA acceleration time (PAAT), pre-ejection time (PET) and RV Ejection time (ET) were measured using 2 d echocardiogram and Doppler. RV mechanics and RV to PA coupling was assessed by RV work (TAPSE X PASP/PAAT) before and after the Potts shunt and at last follow up. 4/8 underwent cardiac catheterization prior to the procedure. N terminal pro-brain natriuretic peptide (NT-proBNP) and Six min walk test was performed prior to the procedure and at follow up. All the patients received dual pulmonary vasodilator therapy with phosphodiesterase 5 inhibitors and endothelin receptor antagonists. In addition, they received antifailure treatment for right heart failure and inhaled nitric oxide whenever required. After initial stabilization, 6/8 underwent surgical POTTS shunt using an interposing Gortex tube graft and 2/8 underwent PDA stenting.
Results: 4/6 patients in the surgical group and 2/2 in the PDA stenting survived the procedure. Both the patients who died had supra-systemic PA pressures and severe right ventricular dysfunction. All the survivors were followed up in the PAH clinic, median duration of follow up was 6 months (3 months – 19 months). 5/6 patients who survived the procedure had significant improvement in functional class (IV vs II, p= 0.02) with decrease in dose and number of pulmonary vasodilators and improvement in RV function and RV to PA coupling. 1 patient had no change in the functional status. TAPSE z score improved from - 4.5 $\pm$ 1.5 to -1.3 $\pm$ 0.5 (p= 0.02) and RV to PA coupling as assessed by RV work improved from 562 $\pm$ 115 to 883 $\pm$ 113 (p= 0.04). NT-Pro BNP decreased from median of 2650 (1126-3400) to 335 (113-446) p= 0.001 at last follow up. There were no deaths in the patients who survived the procedure.
Conclusion:
POTTS shunt can be considered as an interim palliative procedure as a bridge to lung transplant in patients with idiopathic PAH with reasonable success with improvement in functional class and decrease in the requirement for medication.