

POTTS shunt as a palliation for patients with idiopathic PAH.

*Bobhate P., Colaco S., Karande T., Kasar P., Kulkarni S.
Childrens' heart centre, Kokilaben Dhirubai Ambani Hospital, Mumbai, India*

Introduction:

Idiopathic pulmonary artery hypertension (IAPH) is a progressive disease and may become fatal in many patients. Survival of Eisenmenger's syndrome is much better as compared with patients with IAPH. POTTS shunt has been shown to improve functional class and prolonged survival in patients with IAPH

Materials and Methods:

This is a prospective study where 10 patients (age= 9 months-35 years) with PAH on maximal medical therapy, in functional class IV. 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. 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. 6/10 underwent cardiac catheterization prior to the procedure. NT-proBNP and 6MWT was performed prior to the procedure and at follow up. All patients were on maximal medical therapy before the surgery. 8 underwent surgical POTTS shunt and 2 underwent PDA stenting.

Results:

5 patients in the surgical group and 2 in the PDA stenting survived the procedure. All three who died had severe right ventricular dysfunction. All the survivors were followed up in the PAH clinic, median follow up was 6 months. 5/7 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 ± 1.5 to -1.3 ± 0.5 ($p= 0.02$) and RV to PA coupling improved from 562 ± 115 to 883 ± 113 ($p= 0.04$). NT-Pro BNP decreased from 2650 (1126-3400) to 335 (113-446) $p= 0.001$. There were no deaths after discharge.

Conclusion:

POTTS shunt can be considered as an interim palliative procedure in patients with PAH. Surgery at the earliest indication of clinical deterioration and preoperative stabilization is essential.