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Bilateral Lung Transplantation and PostOp VA-ECMO: A Novel Approach for Children with Endstage Pulmonary Arterial Hypertension (PAH)

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INTRODUCTION: Despite improvement in pharmacotherapy, bilateral lung transplantation (BLuTx) or combined heart-lung-Tx (HLuTx) remain the only established treatment options for children with endstage PAH. Although PAH is the second most common indication for BLuTx in children, data on the best perioperative management, pre- and postoperative cardiac function and mid/long-term outcome are lacking. We hypothesized that BLuTx, followed by early extubation and awake VA-ECMO, is associated with excellent short- and mid-term outcomes.
METHODS: Retrospective study on children with PAH who underwent BLuTx at Hannover Medical School from January 2010 to September 2015. Pre-, peri- and postoperative data (demographics, echocardiography, cardiac MRI, CPR, ECMO, survival) were collected and analyzed.
RESULTS: 7 consecutive patients with endstage PAH underwent minimal invasive BLuTx (mean age 13yrs; range 7-16yrs; diagnosis: 5 idiopathic, 1 after d-TGA arterial switch repair, 1 pulmonary capillary hemangiomatosis, i.e. group 1' PH). Average time on HU waiting list was 52days (range 1-130d); 3 patients were resuscitated prior to BLuTx, 2 of which were bridged to BLuTx on VA-ECMO. Intraoperative VA-ECMO or cardiopulmonary bypass was applied in 5 and 2 patients, respectively; 5 patients received scheduled post-BLuTx VA-ECMO support (mean duration 8.4days; range 6-12d). The goal "early extubation and awake VA-ECMO" could be achieved in 4/5 patients. Preoperative echocardiography and cardiac MRI showed severely compromised enlarged right ventricles (RV), i.e. suprasystemic or systemic RV pressure, endsystolic septal shift with LV compression, and pericardial effusion. Hemodynamic compromise rapidly improved after BLuTx, i.e. normalization of RV volumes and systolic function, and fade of LV compression and pericardial effusion. As of September 2015, all 7 patients are alive post-BLuTx, currently with a median survival time of 2.7years (range 0-5yrs).
CONCLUSIONS: BLuTx in children with endstage PAH is associated with encouraging results in our center. Post-op "awake VA-ECMO" might help facilitating RV and LV recovery, reducing perioperative and midterm mortality, and thus allowing better outcomes.