ABSTRACT BOOK

28th International Symposium on

Adult Congenital Heart Disease

'Tackling & Transforming Outcome of Complex CHD'

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FEASIBILITY AND EFFICACY OF NEGATIVE PRESSURE VENTILATION IN THE AMBULATORY FONTAN POPULATION-(FONTAN-CMR) - A PILOT STUDY

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Background: The Fontan operation is the procedure of choice for children born with single ventricle physiology. Although cardiovascular complications are relatively uncommon in childhood following successful Fontan palliation, rates of morbidity and mortality are strikingly high in adulthood and can be attributed to a chronically low cardiac output (CO) state. Treatment options for adults with failing Fontan physiology are extremely limited. While short-term augmentation of CO with negative pressure ventilation (NPV) immediately following Fontan surgery has been documented, the effects of NPV in the ambulatory setting have never been studied. Using a novel, non-invasive, portable ventilator (United Hayek Medical) never before applied to the ambulatory congenital heart disease population, our aim was to explore the safety, feasibility and efficacy of NPV in the adult Fontan population. Hypothesis: We hypothesized that NPV will result in an improvement in CO as measured by cardiovascular magnetic resonance imaging (CMR).

Methods: Primary: To explore the impact of NPV on CO in adults with a Fontan circulation as compared with healthy controls. Secondary: To evaluate safety and tolerability of the device by questionnaire. Material and methods: Inclusion criteria: Adult patients (> 18 years) with lateral tunnel or extracardiac Fontan connections and a dominant left ventricle were screened for inclusion from an existing database. Healthy controls were matched 1:1 based on age and gender. Exclusion criteria: Patients were excluded if they had evidence of failing Fontan physiology (as determined by his/her primary cardiologist), significant valve regurgitation/stenosis, obstruction within the Fontan circuit, resting oxygen saturation 35 kg/m2), chest wall deformities and/or contraindications to CMR imaging. Phase contrast (PC) flow measurements using CMR were completed at multiple locations (ascending aorta, descending aorta, pulmonary arteries, vena cavae and Fontan circuit) at baseline and at various timepoints and pressure settings (as shown in Figure 1). Flow measurements were expressed as L/min/m2. Continuous data within and between groups were analyzed using Wilcoxon Mann-Whitney test.

Results: We enrolled 20 subjects: 10 adult Fontan patients and 10 age-matched healthy controls. We completed CMR studies in 8 Fontan patients and 10 controls (2 CMR studies pending at the time of abstract submission). Patient demographics are presented (Table 1). At baseline, Fontan patients had decreased cardiac index (CI) compared to healthy controls (Figure 2). A brief period of intermittent NPV resulted in significant increase in CI from 2.65 to 3.26 L/min/m2 (23% mean increase) and pulmonary blood flow from 2.15 to 2.72 L/min/m2 (26% mean increase) in the Fontan group as compared to controls (Figures 3 and 4). Patients reported better tolerability of the device in the intermittent as compared to the continuous NPV setting; no significant adverse events were observed during the study.

Conclusions: A significant improvement in CO can be achieved in adult Fontan patients using intermittent NPV in the ambulatory setting. This pilot data require further validation in larger populations of Fontan survivors and the potential impact of enhanced CO on clinical outcomes should be the focus of future studies.