STRONG CHILDREN'S RESEARCH CENTER

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ABSTRACT

Title*: Hemodynamics are not associated with measures of sleep disordered breathing or pulmonary function in patients with Fontan physiology.*

Background: The Fontan procedure is a palliative surgical treatment for single ventricle congenital heart disease. As more children with single ventricle physiology survive into adulthood, comorbidities associated with Fontan physiology, such as obesity and obstructive sleep apnea (OSA), are becoming more recognized. In Fontan physiology, the blood flow returning from both the upper and lower body (through the superior vena cava and inferior vena cava respectively), flows directly into the pulmonary arterial circulation as the single ventricle has been connected to the aorta and is pumping blood to the body leaving no pumping chamber for blood entering the lungs. Because pulmonary blood flow is passive in Fontan circulation, this physiology relies heavily on optimized respiratory function to produce low pulmonary vascular resistance, making it susceptible to the harmful effects of positive pressure ventilation. In order for venous return to flow efficiently into the pulmonary arterial circulation, pulmonary vascular resistance must be low. Any increase in pulmonary vascular pressures either intrinsically, through constriction of the vascular system itself, or extrinsically from increases in thoracic pressures, will decrease blood flow into the pulmonary vasculature. This in turn will cause inadequate preload to the single pumping chamber providing blood flow to the systemic circulation and can also result in back up of blood on the venous side of the body causing increase in pressures in the liver and gastrointestinal vasculature. Obstructive sleep apnea results from upper airway closure during sleep which can lead to intermittent nocturnal hypoxia which can, in turn, induce pulmonary vascular remodeling and increased pulmonary vascular resistance. Many patients who have survived initial Fontan palliation are left with structural chest wall abnormalities as well as diaphragmatic paralysis, both of which reduce lung volumes and are therefore associated with increased risk of airway closure. Treatment of sleep apnea in patients with Fontan physiology is complicated because use of continuous positive airway pressure (CPAP) or BiLevel positive airway pressure (BiPAP) can increase thoracic pressures, decrease venous return and thereby decrease ventricular preload leading to undesired decrements in cardiac output. In contrast, positive airway pressure also functions to reduce afterload which may improve cardiac output. The prevalence and risk factors for obstructive sleep apnea in patients with Fontan physiology are largely unknown.

Objective: This study aims to investigate the occurrence of obstructive sleep apnea and its relationship to ventilatory efficiency, exercise capacity, lung volumes, respiratory muscle strength, Fontan hemodynamics, airway resistance, and obesity in individuals with Fontan palliation.

Methods: This study was a retrospective single-center review of 112 adult patients with Fontan palliation who received care at the University of Rochester Medical Center between January 2016 and December 2022. Patients were identified using the institutional electronic medical records and cardiothoracic surgery (STS) database using ICD 10 & CPT codes. Medical records were reviewed for demographic data, anthropometric data (height, weight, and BMI), past medical and surgical history, Fontan hemodynamics from cardiac catheterization (Venous and arterial saturations, Fontan mean pressure, pulmonary capillary wedge pressure (PCWP, mmHg), ventricular end-diastolic pressure (EDP, mmHg), pulmonary vascular resistance (PVR, Wood Units), cardiac index (CI, L/min/m^2)), chest x-ray for the

presence of diaphragmatic palsy, chest wall deformities (scoliosis, kyphosis, kyphoscoliosis), cardiac MRI or CT (Fontan type, size of the Fontan baffle, ventricular function), pulmonary function tests (PFTs), and sleep study results. Each of the primary investigators had routine access to these charts as patients who have undergone Fontan palliation are typically followed by pediatric specialists until adulthood and then jointly with adult specialists due to the complex nature of the cardio-pulmonary physiology in this patient population.

Results: Lung size (forced vital capacity) is tightly associated with the maximum volume of air released in one second during forced exhalation (FeV1, $p<0.005$) as was anticipated. Increases in pulmonary capillary wedge pressure (PCWP) were associated with decreased cardiac output ($p<0.05$). Body mass index (BMI) was positively associated with PCWP ($p<0.05$) but did not have a statistically significant relationship with either CI or PVR. Overall, measures of cardiac function including PCWP, PVR and CI were not statistically significantly associated with depth of nocturnal desaturation (p=n.s.), duration of time spent with O2 saturations below 90% (p=n.s.) or mean O2 saturations during overnight polysomnography (p=n.s.). Likewise, measures of cardiac function were not associated with obstructive apnea hypopnea index (o-AHI, p=n.s.) or measures of pulmonary function obtained during spirometry (p= n.s.).

Conclusions: In our patient population, fewer patients had undergone either polysomnography or pulmonary function testing than was anticipated. We did not find a statistically significant relationship between Fontan hemodynamics and measures of either nocturnal desaturation or obstructive sleep apnea. Likewise, we did not find a statistically significant relationship between Fontan hemodynamics and pulmonary function obtained during spirometry.