Pulmonary Screening in Subjects after the Fontan Procedure

Deborah R. Liptzin, MD[1], Michael V. Di Maria, MD[2], Adel Younoszai, MD[2], Michael R. Narkewicz, MD[3], Sarah L. Kelly, PsyD[2], Kelly R. Wolfe, PhD[4], and Livia A. Veress, MD[1]

Objectives To review the pulmonary findings of the first 51 patients who presented to our interdisciplinary single-ventricle clinic after undergoing the Fontan procedure.

Study design We performed an Institutional Review Board–approved retrospective review of 51 patients evaluated following the Fontan procedure. Evaluation included history, physical examination, pulmonary function testing, and 6-minute walk. Descriptive statistics were used to describe the population and testing data.

Results Sixty-one percent of the patients had a pulmonary concern raised during the visit. Three patients had plastic bronchitis. Abnormal lung function testing was present in 46% of patients. Two-thirds (66%) of the patients had significant desaturation during the 6-minute walk test. Patients who underwent a fenestrated Fontan procedure and those who underwent unfenestrated Fontan were compared in terms of saturation and 6-minute walk test results. Sleep concerns were present in 45% of the patients.

Conclusions Pulmonary morbidities are common in patients after Fontan surgery and include plastic bronchitis, abnormal lung function, desaturations with walking, and sleep concerns. Abnormal lung function and obstructive sleep apnea may stress the Fontan circuit and may have implications for cognitive and emotional functioning. A pulmonologist involved in the care of patients after Fontan surgery can assist in screening for comorbidities and recommend interventions. *(J Pediatr 2018;■■:■■-■■).*

Patients born with single-ventricle physiology typically undergo staged surgical palliation culminating in the Fontan procedure, which involves the formation of a connection between the inferior vena cava and the pulmonary artery, creating a total cavopulmonary connection with entirely passive flow to the lungs. Contemporary modifications to the Fontan operation include a fenestration between the Fontan conduit and the atria, providing pressure relief in the setting of elevated pulmonary arterial pressures.

Plastic bronchitis is a life-threatening pulmonary morbidity that can occur after the Fontan procedure.[1] One theory is that plastic bronchitis occurs secondary to high lymphatic pressures and/or abnormal lymphatic drainage, leading to the leaking of fibrin-rich lymphatic fluid into the airways, which then solidifies and takes the shape of the airways, forming fibrin casts, which can potentially lead to fatal airway obstruction.[1] In addition to plastic bronchitis, any pulmonary process that increases pulmonary vascular resistance can strain the Fontan circuit and ultimately decrease cardiac output. For that reason, a pulmonologist was included in the interdisciplinary single-ventricle clinic at our institution. We reviewed the pulmonary findings of the first 51 patients who presented to our interdisciplinary outpatient clinic after undergoing the Fontan procedure.

Methods

Our interdisciplinary single-ventricle team formed to address the complex needs of patients with single ventricles comprises a nurse coordinator and pediatric providers from cardiology, gastroenterology/hepatology, pulmonology, psychology, neuropsychology, child life, social work, and nutrition.

On Institutional Review Board approval, a retrospective review was conducted of the first 51 patients who presented to our interdisciplinary single-ventricle outpatient clinic after undergoing the Fontan procedure. Pulmonary evaluation included pulmonary function testing, 6-minute walk, and oxygen saturation at the time of interdisciplinary clinic evaluation. Spirometry, lung volumes, and diffusion capacity tests were performed for patients aged 8 years and older; only spirometry was performed in those aged 4-8 years. A bronchodilator was administered unless a parent or patient refused. Spirometry was performed with the CareFusion MasterScreen system with a SentrySuite interface, and body plethysmography was performed with the CareFusion Vmax System (Vyaire Medical, Chicago, Illinois). Reference

FEV1	Forced expiratory volume in the first second
TLC	Total lung capacity

From the [1]Pediatric Pulmonary Medicine, University of Colorado School of Medicine and Children's Hospital Colorado; [2]Heart Institute, University of Colorado School of Medicine and Children's Hospital Colorado; [3]Digestive Health Institute, Children's Hospital Colorado and Section of Pediatric Gastroenterology, Hepatology and Nutrition, University of Colorado School of Medicine; and [4]Pediatric Neurology, University of Colorado School of Medicine and Children's Hospital Colorado, Aurora, CO

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equations for spirometry included Eigen for children aged 3-5 years,[2] Wang for those aged 6-8 years,[3] and Hankinson for children older than 8 years.[4] Reference equations for lung volumes included Zapletal for children aged 5-18 years[5] and European Community for Steel and Coal for those aged ≥18 years.[6] Diffusion capacity reference equations included Stam for children aged <18 years[7] and European Community for Steel and Coal for those aged ≥18 years.[8] Airflow obstruction was defined as forced expiratory volume in the first second (FEV_1) < 80% predicted, FEV_1/forced vital capacity ratio <80, and/or forced expiratory flow 25%-75% (FEF 25-75) < 65% predicted. Reversibility to bronchodilator was defined as a ≥12% increase in FEV_1 and/or a ≥32% increase in FEF 25-75. Restriction was defined as a total lung capacity (TLC) of <80% or, if only spirometry was performed, in both FVC and FEV_1 <80%. Air-trapping was defined as residual volume/TLC of ≥30. A low diffusion capacity was defined as <80% of predicted when adjusted for alveolar volume. The 6-minute walk test was performed according to the American Thoracic Society's standard protocol.[9] Parents and/or patients were interviewed to identify any sleep concerns, including symptoms consistent with obstructive sleep apnea, such as pauses in breathing or heroic gasps.

Descriptive statistics were used to describe the population and test data. The Wilcoxon rank-sum test was used to compare median oxygen saturation level and distance walked during the 6-minute walk test, and the Fisher exact test was used to compare saturation during the 6-minute walk test, between the patients who had undergone a fenestrated Fontan procedure and those with an unfenestrated Fontan procedure.

Results

Over a 12-month period, 51 patients who had undergone a Fontan procedure were evaluated at our clinic, 45 of whom were seen by a pediatric pulmonologist during this period. Eight patients were already being followed by a pulmonologist outside of this clinic (**Table I**). The median patient age was 10.8 years (range, 3.3-21.3 years). The proportion of males and females was comparable (27 vs 24, respectively). The median home altitude of residence was 1613 m (range, 844-2201 m). The underlying cardiac abnormality leading to the Fontan procedure was hypoplastic left heart syndrome (HLHS) in 31% of the patients (n = 16). Fenestration within the Fontan circuit was present in 43% (n = 22).

Pulmonary disorders diagnosed in this post-Fontan population are summarized in **Table II**. Pulmonary concerns were present in 61% (n = 31) of the patients evaluated in this study and included plastic bronchitis, asthma, other pulmonary function abnormalities, and/or sleep abnormalities.

Three patients (6%) had a previous diagnosis of plastic bronchitis, and 5 patients (10%) had a new or prior diagnosis of asthma. No patients reported recurrent pneumonias, symptoms of aspiration, or complications from diaphragmatic paralysis. Sleep concerns were identified in 45% of patients (n = 23), the majority of whom had clinical symptoms concerning for obstructive sleep apnea (n = 17), including snoring, apneic pauses, and/or heroic gasps during sleep. Six additional patients had various other sleep concerns, including restlessness, poor-quality sleep, crying, bedwetting, and difficulty falling asleep.

Table I. Demographic data for the first 51 patients who presented to our interdisciplinary single-ventricle clinic

Characteristic	Value
Age at visit, y median (range)	10.8 (3.3-21.3)
Sex, n	
Male	27
Female	24
Race, n	
White	36
Other	9
Black	2
Asian	2
Multiple races	2
Ethnicity, n	
Non-Hispanic	29
Hispanic	22
Congenital heart disease, n	
HLHS	16
TA	14
DORV	7
TGA	3
Other	11
Fenestration status, n	
Fenestrated	22
Unfenestrated	29
Home altitude, m, median (range)	1613 (844-2201)

DORV, double-outlet right ventricle; *HLHS*, hypoplastic left heart syndrome; *TA*, tricuspid atresia; *TGA*, transposition of the great arteries.

Pulmonary testing results are presented in **Table III**. Baseline oxygen saturation levels were obtained for 48 patients. The median baseline (room air) oxygen saturation in the clinic was 91% (range, 76%-99%). In the patients with a fenestrated Fontan, the median baseline oxygen saturation was 87.5% (range, 76%-96%). In patients with an unfenestrated Fontan, the median baseline oxygen saturation was 92.5% (range, 80%-96%). Saturation was significantly lower in the patients with a fenestrated Fontan ($P = .002$). Pulmonary function testing was performed in 38 patients; the other patients could not be tested because of young age, significant developmental delays, or scheduling difficulties. The test data showed normal pulmonary function in 8 patients (21%). Airway obstruction and/or air trapping was present in 12 patients (32%). Two patients had reversibility on pulmonary function testing, 1 patient had a history of reversibility on pulmonary function testing, 1 had

Table II. Pulmonary disorders diagnosed in the first 51 patients who presented to our interdisciplinary single-ventricle clinic

Variable	n
Previously evaluated by pulmonologist	6
Sleep concerns	23
Snore/gasp/pause	17
Other	6
Asthma	5
Plastic bronchitis	3

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