The focus of this case study is a twenty-four-year-old, Caucasian, female diagnosed with Idiopathic Pulmonary Hypertension (IPHTN), type II Diabetes Mellitus, asthma, migraines, and severe protein malnutrition. The patient had proceeded with surgical treatment for IPHTN—a progressive and debilitating disease, via a bilateral cadaveric transplant more than six years before at a pediatric transplant center. Unfortunately, her post-transplant trajectory was complicated by an unforeseen need to temporarily halt the usage of her anti-rejection medications. The temporary hiatus from her transplant medications resulted in an irreversible condition known as Bronchiolitis Obliterans Syndrome (BOS). This life-threatening pathology led her to seek medical consultation at another adult regional transplant center located in Philadelphia.

On April 8, 2016, the patient was transferred from that regional transplant center to Kindred Hospital of South Philadelphia for physical rehabilitation and nutritional support in the hope of re-qualifying her for yet another exploration into lung re-transplantation. Upon transfer to our facility, the patient was evaluated and presented as cachexic, anxious, and notably dyspneic. Our goal was to ramp up the patient’s physical conditioning and address her muscle-wasting syndrome.

The elevation in oxygen consumption related to patients with profound lung pathology has a deleterious effect on the body’s ability to retain muscle mass. In addition, the associative physiologic finding of an increased Respiratory Quotient (RQ)—spending a significant amount of energy on respiratory effort—prevents these patients from actively participating in physical rehabilitation. Fortunately, new advances in ventilation technology provide hope for many patients that were once delegated to a sedentary and isolated lifestyle. Breathe Technologies, Inc. offered just that solution for our patient to participate in a trial with their proprietary Non-Invasive Open Ventilation (NIOV) System to decrease her overall work of breathing (WOB) and RQ. The results were nothing less than remarkable. The patient’s former intractable BMI of 13.70 and weight of 72.70 pounds was improved notably to a BMI of 16.90 and a weight of 89.60 pounds, a gain of 16.9 pounds, within a three week time period. Another benefit gained from using the NIOV System was the significant improvement in her endurance tolerance. Prior to her transfer and using the NIOV System, the patient was walking less than 200 feet with notable oxygen desaturations into the mid-70 percentages. Now, while using the NIOV System, the patient was energized to complete a six-minute walk achieving more than 1,250 feet without any notable desaturations.

These milestone accomplishments are compelling predictors of successful lung transplantation in the first post operative year. Clinical research has shown that being underweight is an independent risk factor in morbidity after lung transplantation. In addition, the improved functional ability of this patient’s six minute walk distance may represent a positive impactful factor in diminishing post-operative risk factors.

The NIOV System afforded this young woman with new found opportunities to pursue activities of daily living that we all too often take for granted. Using the NIOV System throughout the day renewed her confidence and determination to successfully cope with physical endurance challenges ahead. Supported by Breathe Technologies, this young woman was able to transition back to her home, which was more than one hundred miles from our hospital. Now she is surrounded by her loved ones with a sense of hope that her next visit to the hospital will be to receive her new lungs.

References