

Bridging Patients To Lung Transplant – A Cystic Fibrosis Case Study

Chris Landon MD FAAP, FCCP, CMD

Case Study Background

When it comes to cystic fibrosis, the damage to a human body is devastating. An estimated 30,000 children and adults in the United States have this genetic disease.

Everyone takes breathing for granted — imagine taking a straw and trying to breathe through it for an entire day and you get the idea of the impact of cystic fibrosis.

Patients suffer a variety of impacts, including shortness of breath and increased oxygen need, which severely decreases a person's ability to do even the most basic forms of physical activity. The condition leads to a variety of wasting of vital muscle groups — respiratory muscles, postural or trunk muscles, arm and leg muscles, and cardiovascular deconditioning.

But cystic fibrosis doesn't just erode the body physically — a patient's mind suffers as well because they are no longer able to do even the most basic things. Climbing a couple of steps to get into their home becomes impossible without gasping for air. Standing and doing the dishes can become a catastrophic experience because this breathlessness leads to intense fear or anxiety.

Lung transplantation is possible for patients with end stage cystic fibrosis, but not everyone is healthy enough to qualify or even survive such a major surgery.

Without intervention, a poor outcome is inevitable. That's why the team of professionals at the Children's Hospital Los Angeles affiliated Cystic Fibrosis Care Center at Ventura County Medical Center all worked together to conduct a case study to see how the Breathe Technologies' NIOV System could improve the outcome of a patient with this disease.

The Patient

J was chosen from the Adult Cystic Fibrosis Clinic. He is 32, works as a banker and had recently gotten married. His life-long physical activity was playing ice hockey, and he also enjoyed free weights and jogging on a treadmill, activities he was unable to continue due to his condition.

Chris Landon MD FAAP, FCCP, CMD is Clinical Associate Professor of Pediatrics Keck-USC School of Medicine, Director of Pediatrics, Ventura County Medical Center and Center Director of the Children's Hospital Los Angeles affiliated Cystic Fibrosis Care Center Clinic at Ventura County Medical Center.

J was on the transplant list when chosen for the case study, but without an improvement in his condition he was not healthy enough to be able to survive such a major surgery.

J's end stage lung disease was marked by FEV1 of 16%, every six-week three-week courses of antibiotics, and the placement of a gastrostomy tube to maintain weight. Despite nasal cannula oxygen at 4 LPM, J was still unable to participate in his exercise program. Serial six-minute walk tests had been performed with each clinic visit. He was having difficulty ascending stairs at home despite the use of oxygen. The St George's Respiratory Questionnaire Symptom Score 85 (normal 12), Activity Score 92 (normal 9), Impact Score 66 (normal 9), and Total 77 (normal 2) was consistent with the devastating impact on his life.

Treatment Plan

The medical team developed a single subject design for J consisting of Baseline and Intervention with the Breathe Technologies NIOV System. NIOV consists of a one-pound portable device that can be mounted on a belt clip, making it easy for J to carry with him. A four-hour battery duration allows for extended trips. J was titrated by delivering volume of 100 mL at rest and 140 mL with exercise. The NIOV System detects a J's spontaneous breathing via sensor ports located in the nasal pillow interface and delivers synchronized augmented volumes of ambient air and oxygen at rates of up to 40 breaths per minute.

Testing Methods

At baseline a six-minute walk test was performed with and without the NIOV System and repeated at Day 30. Primary endpoint was the 6 Minute Walk Test distance and Borg score.

The Results

Using the NIOV System, J's life has seen a remarkable turnaround.

J was able to participate in the physical activity required to keep enough muscle mass and stamina to survive transplant surgery. At that point during our study, all J needed was a healthy set of lungs in order to move forward with the transplant.

J was also strong enough to go on a belated honeymoon.

By day 30 of using the NIOV System, J's condition had greatly improved and his rating on the transplant inclusion criteria scale was upgraded. J noted a ventilator synchrony problem with

movement and unexpected ventilator assistance while driving. Adjustments were sufficient to make the intervention tolerable.

For the 6 Minute Walk Test, at baseline J was only able to cover 980 feet with nasal cannula oxygen alone, but on that first day with the NIOV System he was able to increase that number to 1,050 feet. After 30 days of exercise augmented by the NIOV System, J was able to walk 1,050 feet with nasal cannula and 1,110 feet with the NIOV System.

J's Borg scores greatly improved as well after 30 days of using the NIOV System, dropping from a Borg baseline of 6.5 after 4 minutes to a score of 2, and a Borg baseline of 4 after 8 minutes to zero.

J Updates His Progress

Since the case study was conducted, J has updated the medical team on his daily activities, saying that the NIOV System is an essential part of his exercise routine. Here is what is involved in his own words:

“As I continue to deteriorate, the NIOV has had a bigger and bigger impact. Right now if I try to go on the elliptical while on the regular cannula at 6LPM, I can last about 20 seconds before my O₂ drops to below 88 and I am completely out of breath. While on the NIOV, I have it at 180 mL, I have built up to going 4 minutes without stopping, and I stop because of fatigue rather than because of my O₂ dropping (O₂ only takes a slight dip to about 90/91 by the time I need to stop). I go on the elliptical every single day for at least 3 minutes. I work out with weights for about 5 minutes, 5 times a week. It doesn't sound like much, but without the NIOV I wouldn't be able to do anything at all, and I would probably be in the hospital by now - too sick for a transplant at this point.”

Then came the most dramatic update – with J writing with incredible news that due to the NIOV System he was healthy enough to receive that transplant.

“I am emailing you because last night I got home from my lung transplant after being in the hospital for 10 days. I have been feeling amazing, and was very lucky to have a speedy recovery, was able to walk within a couple hours after waking up, and was walking 2 miles per day after just a couple days. I probably won't be needing the NIOV device anymore, and I cannot tell you enough on how much of an absolute life-changing, enormous help that was. With that device I was able to go on the elliptical for 5 minutes at a time every day, and just do more. With the straight oxygen at 6 LPM, I could barely walk down the hall. Even the surgeon told me that he could not believe I was able to exercise, let alone stand with how bad my lungs were (my FEV₁ was down to 10%). Because I had the device, I was able to stay strong, which was a tremendous part of me recovering so well, and also keeping me alive that much longer.”

More About The Device

When the patient inhales, a set volume of oxygen gas is delivered using the NIOV System — all customized for that patient. Volume delivery settings ranging from 50 mL to 250 mL can be quickly programmed to 3 levels of patient activity — low, medium and high. In addition to the volume of oxygen gas that is preset, ambient air is also entrained through two entrainment ports located on the interface to increase the total volume and properly ventilate patients.