Slowing, Stabilizing, Improving Lung Disease

Advanced lung disease (ALD) has a high mortality rate but is yielding more to management as a result of improved experience with medications and better bronchoscopic procedures. Chronic, progressive respiratory failure refractory to treatment that has progressed to the point where the patient is severely disabled or faces death from the disease in a short period of time is the late stage of various obstructive or interstitial lung conditions.

"Unfortunately, some patients progress to this point of ALD, in spite of best standard medical practice. Generally, ALD is present when the lung function has dropped below about a third of normal function. At that point, most patients require oxygen and are quite limited," said Virtua pulmonologist Patrick Mulhall, MD. "But in our advanced lung disease program, we find that improved scoping procedures and better application of more varied antifibrotic and immunosuppressant medications are making a difference."

Fighting Progression, Preserving Lung Function

Categories of pulmonary conditions that lead to ALD are interstitial (restrictive) lung diseases—a heterogeneous group of fibro-inflammatory disorders causing destructive deposition of collagen in lung parenchyma (e.g., idiopathic pulmonary fibrosis, lupus, sarcoidosis)—and obstructive lung diseases (e.g. COPD, asthma). CT imaging and bronchoscopy for lavage and biopsy now replace open lung sampling.

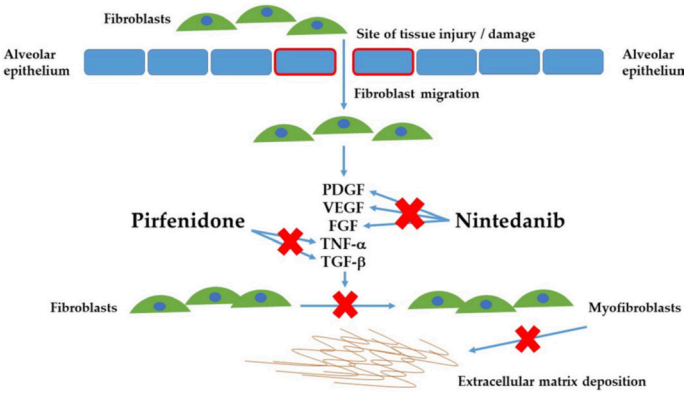
"And instead of video-assisted wedge resection for lung nodules, we now use robotic-assisted bronchoscopic cryobiopsy with radiologic guidance, which permits us to extract nodule tissue with near-certain precision to differentiate cancerous lesions from ALD," said Virtua interventional pulmonologist Mark Weir, MB ChB.

ALD specialists now use a variety of oral and IV anti-rejection medications (with steroids) and anti-scarring medicines to treat fibrotic lung disease. Used alone or in combination, these drugs can stabilize and even improve forced vital capacity and add years to survival. Mycophenolate, rituximab (a monoclonal antibody), and tacrolimus decrease lymphocyte and inflammatory cytokine release to protect the lungs from ongoing damage and inflammation. The antifibrotics nintedanib and pirfenidone, now available in generic form, decrease growth factor signaling to reduce collagen synthesis and extracellular matrix production *(see figure)*. The Virtua pulmonologists can titrate ALD patients to their tolerated doses.

Managing Around the Lung Transplant Option

For patients with a very low volume of functioning lung, such as those with advanced COPD, Virtua is the only center in South Jersey that offers a new and preferred alternative to surgical lung reduction. Implantable one-way Zephyr® valves help to isolate unhealthy tissue and push breathing capacity to healthier areas of the lungs *(see page 2)*.

The staff of Virtua's advanced lung disease program can also manage patients with the goal of avoiding a lung transplant. At the same time, they test and identify those for whom transplant is required, help to optimize their care leading into transplant, and manage their immunosuppression and other care post-transplant.



Source: *Journal of Clinical Medicine*

In the treatment of fibrotic lung disease, nintedanib and pirfenidone bind intracellularly to key receptors to inhibit fibroblast proliferation, migration, and transformation into myofibroblasts.

For patient appointments, call 888-VIRTUA-3 (888-847-8823).

Virtua Health

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Valves Shunt Air to Healthier Lung, Improve QOL

Case review: With an extensive history of smoking, musician 76-year-old Frank Knox of Ocean County suffered from very severe COPD. A virus hospitalized him in 2007, further weakening his breathing capacity, and repeated bouts of pneumonia in 2023 additionally injured his lungs. His life expectancy without a lung transplant was short. A friend of his learned about the Zephyr® valve. Knox's pulmonologist was unfamiliar with the valve, but referred him to Virtua, the only health system in South Jersey to offer the procedure.

When Knox arrived at Virtua in April of this year, only 17% of his lung capacity remained. He required three liters of oxygen per day and was mostly housebound. CT imaging showed that his left upper lobe was most deteriorated. The Virtua team implanted seven Zephyr valves bronchoscopically in the secondary and tertiary bronchi of this lobe. The tiny, one-way valves permit air to exit but not enter the treated lobe, which then deflates as airflow is redirected to the more functional lobes of the lung.

Knox recovered in the ICU, particularly to guard against rupture and pneumothorax of the lower left lobe as that lobe hyperinflates with new airflow. After three nights, he walked out of the hospital to applause, no longer wheelchair bound.

Within one month of discharge, the treatment had eased Knox's shortness of breath enough for him to take a vacation cruise. Today, his level of activity permits him to engage in social events and play with his granddaughter. He currently uses only one liter of oxygen per day. He's even singing a bit, albeit in the car rather than on stage.

"I was housebound. I couldn't even walk to the mailbox," he said. "Now I feel so much younger and healthier. It's unbelievable!"

Zephyr endobronchial valve treatment shrinks damaged areas of the lung as it diverts air to the healthier lobes of the lung, which now have more space in the chest to expand and function. The pulmonary specialists can remove and replace the valves, or treat a second lobe in the same way, if needed, in a subsequent procedure.

"Zephyr valves can potentially help patients delay or even avoid lung transplant, thus providing an additional option to transplant, which is a quite invasive and high-risk operation. But it also doesn't exclude them from transplant, and can serve as a bridging therapy to transplant in some cases," said Virtua interventional pulmonologist Syed Riaz, MD. "We are glad to be able to offer this option, because even a little improvement in breathing is significant for these patients. We assess the benefit objectively with imaging and exercise capacity testing, but the patient's subjective improvement —of feeling better and breathing better—is the main goal."

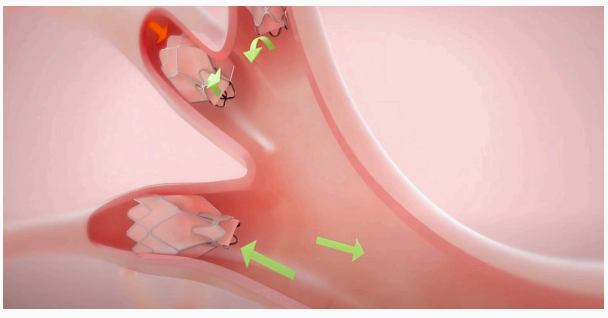

Illustration courtesy of Pulmonx

When medical management alone isn't enough for end-stage COPD, minimally invasive Zephyr valve implantation can occlude damaged areas of the lung and redirect airflow to healthier portions of the lung.