



The Spirogram

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Doctor I have Idiopathic Pulmonary What?

Yolanda Mageto, M.D.



Idiopathic pulmonary what? What is it? Can it be cured? How did I get it? Is it contagious? Will my family need to be checked? Why wasn't I diagnosed earlier? These are the typical questions patients often ask upon hearing that they have been diagnosed with Idiopathic pulmonary fibrosis.

Idiopathic Pulmonary Fibrosis: What is it?

Idiopathic pulmonary fibrosis, otherwise known as IPF, is a progressive scarring of the lung over time. When the lungs become more scarred they are unable to expand and the patient develops a restrictive pattern on pulmonary function testing. In the majority of patients insidious shortness of breath or dyspnea is the initial symptom and often is unnoticed until a significant amount of scarring has occurred. The reason many people do not seek medical attention earlier because this disease typically affects those over 50 and most attribute their symptoms to aging. Some patients may present with a dry hacking cough for a number of months and are misdiagnosed as having heart failure; others might initially develop a severe pneumonia that causes them to seek medical help.

What causes the disease?

No one knows for certain. Idiopathic pulmonary fibrosis belongs to a small group known as the idiopathic interstitial pneumonias of which IPF is the most common. Recent research suggests that the lung itself is exposed to a variety of insults over time. Whenever the lung is injured by something we inhale there are numerous events/reactions that occur leading to self healing using the bodies own immune system. However, in pulmonary fibrosis it progresses to inflammation and then on to a fibrotic state somewhat like a scratched CD or LP. Because of this scarring, oxygen is unable to cross from the lung into the blood stream and patients typically require oxygen.

Why wasn't I diagnosed earlier?

Unfortunately until roughly 5-7 years ago most people thought this disease was rare believing that it occurred in only 7/100,000 people. More recent estimates suggest that there are roughly 80,000 cases in the US and 30,000 new cases a year. IPF is most often confused with congestive heart failure because on physical examination patients have crackles or rales (which sound like Velcro) that are similar to the sounds one hears with congestive heart failure. Thus many patients are evaluated initially by a cardiologist delaying the time of referral to a pulmonologist. Other possible disorders that present in a similar fashion include patients with collagen vascular disorders such as

rheumatoid arthritis or scleroderma, chronic hypersensitivity pneumonitis or chronic sarcoidosis. Other advancements that help with making an earlier diagnosis include the recognition by radiologists and physicians of some of the early subtle changes on a chest radiograph. In fact many patients have chest radiograph changes long before they have symptoms.

How is Idiopathic Pulmonary Fibrosis diagnosed?

The gold standard for diagnosis remains an open lung biopsy which involves having several small pieces of lung tissue removed and sent to the pathologist. Over the last few years a number of studies have been done to see if a group of clinical factors and tests can be used to accurately make the diagnosis of IPF without an open lung biopsy. Results suggest, by using a combination of CT scans, pulmonary function tests, blood work and clinical history that an experienced pulmonologist can make an accurate diagnosis. Patients who are sent for biopsy are those with atypical presentations who don't fit the classic clinical picture.

Is IPF contagious? Is my family at risk?

IPF is not contagious however your family may be at risk since there is an entity known as familial idiopathic pulmonary fibrosis. There are number of researchers searching for a pulmonary fibrosis gene or genes to see if those at risk can be identified early. If one has a diagnosis of IPF or has a family member with Idiopathic Pulmonary Fibrosis it might be advisable to take a family history for lung disease but the majority of idiopathic pulmonary fibrosis is sporadic and does not run in families. There are some risk factors however and these include, smoking, male gender, age, ethnicity (more common in Caucasian), and occupations such as: farming, hairdressing, raising birds, stone cutting/polishing, exposure to metal dusts, vegetable and animal dusts.

Can it be cured?

IPF patients are typically managed with a variety of agents; these are typically an immunosuppressive agent and low dose steroids. The only real cure is a lung transplant which is fraught with risks and problems. The age cutoff for a transplant at most centers is 65 years young.

Today there are a number of clinical trials all over the United States and world testing various agents to treat the disease. The Vermont Lung Center is participating in a number of these trials testing new treatments for people with IPF. We hope that these trials will find new and effective treatments for this devastating disease.

Interested in Volunteering?

Things to know.

- 1) The Vermont Lung Center staff is responsible for making sure you know what is expected of you in regards to the study.
- 2) Once the study is explained to you, you will be asked to read and sign an "Informed Consent". This form is designed to explain everything you need to know about the study.
- 3) Studies may be therapeutic (involving observation of lung function). However The Vermont Lung Center can make no claims that your involvement in a research study will improve your condition.
- 4) Compensation may or may not be provided to you for your involvement in a study. If compensation is provided, it is meant to cover your time and expenses incurred—it does not constitute employment.

If you are interested in finding out more about volunteering for a research study, please call us at (802) 847-2193



New Face at the Vermont Lung Center

Julie Martin



What do you do at the Vermont Lung Center?

I work as dietitian for research studies at the Vermont Lung Center and I also coordinate research studies in the medical intensive care unit at Fletcher Allen Health Care.

Where did you grow up?

I was born in California but I spent most of my childhood in Portland Oregon.

Where did you go to school?

I went to high school in Oregon and graduated from Oregon State University. My dietetic internship was completed at the University of Wisconsin. I earned a Master's degree in 2000 from the University of Vermont.

Why did you choose to live in Vermont?

I moved to Vermont 17 years ago because it looked like a great place to raise my children. My two sons are now in high school in South Burlington.

What is your favorite thing about working in research?

It is important for me to feel my job has a purpose in helping people. In our research, we are working toward developing or testing new treatments to hopefully improve lung function and health. It is satisfying to think that our patients or future patients will benefit from the work we are doing.



Clinical Trial of Adult Stem Cells for COPD

Daniel Weiss, M.D., Ph.D.



Stem cells have been prominent in recent news as potential therapeutic approaches for many diseases including lung diseases. Armed with the capacity to limitlessly copy themselves and grow into any type of cell in the body, they function as the body's cellular repair shop.

While embryonic stem cells have been the focus of heated nationwide debate since 1998, many scientists have been examining the potential therapeutic possibilities of controversy-free and promising alternatives – adult stem cells and umbilical cord blood stem cells.

Certain types of adult stem cells derived from the bone marrow, notably mesenchymal stem cells (MSCs), appear to have potent anti-inflammatory properties. This occurs even if the cells don't participate in re-growth of damaged organs, and have been the basis of several successful clinical trials in heart and other diseases. Importantly, the MSCs appear to be well tolerated without any significant serious side effects in the clinical trials to date.

Given this, a new trial has opened investigating whether MSCs will effective for COPD. This is a multicenter, placebo-controlled study to evaluate the safety and efficacy of PROCHYMALTM

for the treatment of subjects with moderate to severe COPD. PROCHYMALTM is the company's trade name for their MSCs. This initial investigation focuses on the safety and possible efficacy of using MSCs for treatment of COPD. Eligible patients will receive 4 infusions of stem cells or of placebo over a four month period. Follow-up will occur over a two year period from the date of the 1st infusion.

The Vermont Lung Center is one of five participating institutions in this study and we're looking to recruit 15 patients. The minimum qualifications are: 1) Age 40-80 years with a diagnosis of Chronic Obstructive Pulmonary Disease; 2) Have a current or former smoking history; 3) have no other significant lung diseases including asthma, lung cancer, pulmonary fibrosis, or tuberculosis. If you are interested or have any further questions, please call Dan Weiss at 656-8925 or Stephanie Burns at 847-2103 You can also read about the trial on the FDA's website www.clinicaltrials.gov.

You may come across uses of stem cells for treatment of COPD originating from Tijuana, Buenos Aires, or other locations. Please DO NOT participate in these or any other trial that has not been approved by the FDA and is not being run by an accredited university or medical center. We will keep you informed about legitimate trials using stem cells when they occur.

List of Current VLC Studies

ASTHMA

Study of Acid Reflux in Children with Asthma (SARCA)

Primary Investigator: Charles Irvin, Ph.D., Director, Vermont Lung Center
 Coordinator: Stephanie Burns
 Who: Children age 6-17 with asthma who do not have heartburn
 What: 9 visits over 7 months
 Compensation: up to \$550

Study of the Impact of Body Mass Index on Asthma

Primary Investigator: Anne Dixon, M.D., Director of Clinical Research
 Coordinator: Laurianne Griffes
 Who: Premenopausal women with asthma and without asthma who have a Body Mass Index of 35-50
 What: 1 to 2 visits
 Compensation: up to \$75

Weight Loss and Asthma

Primary Investigator: Anne Dixon, M.D., Director of Clinical Research
 Coordinator: Laurianne Griffes
 Who: People with asthma and without asthma undergoing gastric bypass or laparoscopic banding surgery
 What: Asthmatics- 10 visits over 12 months; Non-Asthmatics- 4 visits over 12 months
 Compensation: up to \$775 for asthmatics, up to \$250 for people without asthma

Asthma Exacerbations: Physiology, Upper Airway and Fibrin

Primary Investigator: Charles Irvin, Ph.D., Director, Vermont Lung Center
 Coordinator: Sherburn Lang
 Who: People with stable asthma and people without asthma
 What: 2 Visits, each lasting about 3 hours
 Compensation: up to \$225

Forced Oscillation Mechanics in Mild Asthmatics

Primary Investigator: Lennart K.A. Lundblad, Ph.D.
 Coordinator: Sherburn Lang
 Who: People with mild asthma and people without asthma
 What: 3 visits
 Compensation: up to \$15

Role of Leukotrienes and Adenosine in Hyperpnea-Induced Bronchospasm Determined by Dynamic Analysis of Exhaled Breath Condensate

Primary Investigator: John Morrison, D.O.
 Coordinator: Joan Lippmann

Who: People with physician-diagnosed **Exercise-Induced Asthma**
 What: 2 visits
 Compensation: up to \$100

CYSTIC FIBROSIS

Comparison of Standard Tobramycin Inhalation Solution to the new Experimental Tobramycin Inhalation Powder in Cystic Fibrosis

Primary Investigator: Thomas Lahiri, M.D.; Laurie Whittaker, M.D.
 Coordinator: Stephanie Burns
 Who: People with Cystic Fibrosis
 What: 9 visits
 Compensation: up to \$340

CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)

A Phase II, Multicenter, Randomized, Double-blind, Placebo-controlled study to Evaluate the Safety and Efficacy of PROCHYMAL (ex vivo cultured adult human mesenchymal stem cells) Intravenous Infusion for the Treatment of Subjects with Moderate to Severe Chronic Obstructive Pulmonary Disease (COPD)

Primary Investigator: Daniel J. Weiss, M.D., Ph.D.
 Coordinator: Stephanie Burns
 Who: Men and Women 40-80 years of age with moderate to severe COPD
 What: 9 visits over 25 months
 Compensation: \$50 per visit

IDIOPATHIC PULMONARY FIBROSIS (IPF)

Effects of Bosentan on Morbidity and Mortality in Patients with Idiopathic Pulmonary Fibrosis - a Multi Center, Double-Blind, Randomized, Placebo-Controlled, Parallel Group, Event-Driven, Group Sequential, Phase III Study.

Primary Investigator: Gerald Davis, M.D.
 Coordinator: Joan Lippmann
 Who: People with Idiopathic Pulmonary Fibrosis
 What: 2 visits within 4 weeks, then every 4 months visits and monthly laboratory tests.
 Compensation: Travel reimbursement for travel of 20 or more miles.

For more information on these studies,
 please visit our website @
www.vermontlung.org

The Vermont Lung Center is affiliated with the following organizations:



Ask Dr. Charlie

Charles G. Irvin, PhD



What the warning signs of an asthma attack?

There are numerous signs but some of the more important and obvious are: 1.) wheezing both breathing in and out, 2.) coughing that will not stop, 3.) chest and neck muscle tightening 4.) difficulty talking.

I was given an asthma inhaler by my doctor but it doesn't stop my wheezing. Am I using it wrong?

Inhalers especially the meter dose type (liquid type in a small tin canister) are frequently not used properly. Review the use of all your asthma medications with each visit to a health care provider.

The Vermont Lung Center is supported in part by the following organizations:



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We're on the web:
www.vermontlung.org

Fish Oil

Renee Stapleton, M.D.



Fish oils have recently attracted much attention for the health benefits attributed to the omega-3 fatty acids they contain. These omega-3 fats are eicosapentaenoic acid (EPA) and docosahexaenoic acid (DHA). They are essential fatty acids, meaning that humans must consume them because we cannot make them in our bodies. We can ingest fish oil either by eating fish high in omega-3 fats or by taking supplements.

EPA and DHA work to counteract inflammation throughout the body. They have been shown to help in a variety of illnesses including cardiovascular disease, high blood pressure, high triglycerides, and arthritis. Despite the long list of positive effects from fish oils, the vast majority of people in North America, South America, and some parts of Europe are deficient in omega-3 fatty acids. For this reason, many dietitians and health care providers recommend fish oil supplements.

Fish oils play a role in brain function. Studies have shown that low levels of the omega-3 fatty acids are associated with depression and Alzheimer's disease. Higher levels of EPA and DHA are also associated with lower rates of depression and suicide. Another study using mice found that a diet high in omega-3 fats helped prevent the development of Parkinson's disease.

Fish oils are also beneficial to the cardiovascular system. Unlike saturated fats, fish oils protect against heart disease rather than contribute to its development. Omega-3 fatty acids can lower cholesterol levels and blood pressure, and they prevent blood clots. Research shows that fish oils may also prevent abnormal heart rhythms, sudden death from heart attack, and strokes. The American Heart Association recommends the consumption of 1 gram of fish oil daily, preferably by eating fish, for patients with heart disease. The US National Institutes of Health also recommends fish oil for high triglycerides (a type of fat in the blood), prevention of cardiovascular disease, and high blood pressure.

In addition to its positive effects on the brain and cardiovascular health, fish oil has also been shown to reduce pain and inflammation. It is effective in helping to ease the pain of arthritis and colitis (inflammation of the colon), and fish oil supplements may reduce the need for pain medications in patients who suffer from inflammatory conditions. Some evidence also suggests that fish oils might help to prevent or delay the development of breast and colon cancer.

Little is known about the effects of fish oil in lung disease. Researchers at the Vermont Lung Center are currently investigating fish oil to treat critically ill patients with a lung disease called the acute respiratory distress syndrome. Results of this study may lead to a new treatment for this often fatal disease.

