

Transplants – A Treatment Option to Consider

Transplant, PF Doctors Share Insight, Opinion

Lung transplantation is not an option all patients are willing to choose nor is transplant an option available for every patient. But, physicians agree that it is the only life-saving/life-extending option currently available and should be considered immediately upon PF diagnosis.

Keith Meyer, M.D. from the University of Wisconsin School of Medicine and Public Health, generated a questionnaire along with colleagues at the Mayo Clinic to assess physician attitudes toward diagnosis and treatment of PF. Their findings showed support for transplantation for PF patients and were accepted for publication in the peer-review journal *Respiratory Medicine*.

“The majority of pulmonologists who responded to our survey perceive lung transplantation as the best therapeutic option for patients with PF,” Dr. Meyer said. “A majority of respondents (61 percent) felt that lung transplantation represents the only effective therapy for PF, and 86 percent refer their PF patients to lung transplant centers.”

Thanks to the new lung allocation system - the organ transplant program that governs how patients are listed for transplant - PF patients receive priority on the waiting list due their diagnosis and the severity of illness. At many transplant centers nationwide, PF patients make up 50 percent or more of the patients awaiting lung transplantation.

Survival rates of lung transplant patients post transplant is increasing with one-year survival rates now climbing to 80 percent.

“We’d love for it to be 100 percent,” said Steven Nathan, M.D., medical director of the Advanced Lung Disease and Lung Transplant Program at Inova Fairfax Hospital (Fairfax, VA) whose center’s one year-survival rate surpasses the national average at 85 percent. “It is a gamble, but the time to take the gamble is when the greater gamble is not having the transplant. In the context of PF, it is an acceptable alternative.”

According to Dr. Nathan, even PF patients who seem to be early in the course of disease with few or no symptoms should be considered for transplant. “Even if you’re totally asymptomatic, no breathing issues, no cough, no nothing, you need to be seen in a transplant center and be evaluated.”

Jeffrey Golden, M.D., Medical Director of Lung Transplantation and co-Director of the Interstitial Lung Clinic at the University of California, San Francisco, says even patients who might not think they qualify for transplant should ask their doctor about a transplant evaluation. “Patients should tell their doctor they want to be evaluated for transplant. We just operated on three patients with coronary artery disease (without a functional problem) that had been successfully treated for it,” he said.



Steven Nathan, M.D.

*Medical Director of Advanced Lung Disease and Lung Transplant Program
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Keith C. Meyer, M.D.

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Jeffrey A. Golden, M.D.

Professor of Clinical Medicine, Director of the Bronchoscopy Service, Medical Director of Lung Transplantation, and co-Director of the Interstitial Lung Clinic, University of California San Francisco

“The course of PF is so unpredictable that you never know when you’re going to take a turn and need a transplant.”

– Steven Nathan, M.D.

Lung Allocation Score System Saving PF Patient Lives

Recently, the CPF took a look at the Lung Allocation Scoring system, the organ transplant program that governs how patients are listed for transplant, how they place in priority of transplant, as well as how long a patient will wait for new lung(s). We asked three of the country's top PF and transplant experts about the new system, its pros, its cons and why PF patients are being transplanted more than ever, as well as why the system provides hope that was almost non-existent for PF patients in the past.

For more information on the LAS scoring system, visit www.unos.org or contact your physician.

Just a few short years ago, more than a third of PF patients awaiting a life-saving lung transplant didn't receive one in time. Today, the situation for PF patients on the transplant list is much improved and more PF patients than ever are being transplanted and surviving, thanks to the new lung allocation system (LAS) implemented by the Organ Procurement Transplantation Network (OPTN). OPTN is a unified transplant network established by the U.S. Congress to increase the effectiveness of organ sharing and equity in the national system of organ allocation, set up a new program for lung transplantation in 2005.

THE LAS system comprises of a set of precise calculations to determine a patient's listing for transplant (to view the LAS system and calculator, see www.unos.org/resources/frm_LAS_Calculator.asp).

Under the LAS system, patients are now "scored" based on their severity of illness and the likelihood of surviving transplantation, unlike the previous allocation system, which was based strictly on time on the transplant waiting list. Patients with PF generally receive a higher initial score because of the severity

of their disease.

"It represents a real advance in what we're doing," said Steven Nathan, M.D., medical director of the Advanced Lung Disease and Lung Transplant Program at Inova Fairfax Hospital (Fairfax, VA). "The sickest patients [now] get to the top of the list, as opposed to the previous system that was first come, first served."

The United Network of Organ Sharing (UNOS), defines the LAS as a prioritization of waiting list candidates based on a combination of waitlist urgency and post-transplant survival. UNOS administers the OPTN under contract with the Health Resources and Services Administration of the U.S. Department of Health and Human Services.

In this context, according to UNOS, waitlist urgency is defined as what is expected to happen to a candidate in the next year, given their characteristics, if they don't receive a transplant. Post-transplant survival is defined as what is expected to happen to a candidate, given their characteristics, in the first year after a transplant if they do receive the transplant.

"Now, we know this system really helps PF patients. We can put people on the list with severe PF," said Jeffrey Golden, M.D., Medical Director of Lung Transplantation and co-Director of the Interstitial Lung Clinic at the University of California, San Francisco. Before the new system, Dr. Golden said, some PF patients could rarely wait the length of time necessary to be transplanted, so it was futile to even list them.

With the new LAS, PF patients, more often than not, score high because of the diagnosis and the severity of the condition. The higher the score, the more likely a patient is to be transplanted sooner rather than later. In the past, though PF patients received a "90-day credit" in which they received credit for 90 days on the list on the first day they were listed, they were in the diagnostic category most at risk for dying and more than 30 percent of them died before receiving a donor lung.

"The new system also seems to have made organ placement (with various transplant centers) work better and it seems to have decreased the incidence of death on the waitlist," said Keith C. Meyer,

M.D., MS, Medical Director of Lung Transplantation at the University of Wisconsin School of Medicine and Public Health (Madison, WI).

Even though the new LAS system seems to be working well and PF patients are largely surviving long enough to be transplanted, some experts are concerned their severity of illness could affect survival outcomes. "Right now, patients are more ill and they are being transplanted. At least for now, even though they are sicker, their outcomes a year from transplant are the same [as less severely ill patients]," said Dr. Golden. "One worry is that if you transplant sicker patients, they may not survive as long as others. Published data show that is not the case right now."

There is slight concern that the LAS may even favor PF patients too much. "As a matter of fact, the new system, some would opine, has tilted too much toward patients with PF and away from patients with other indications," said Dr. Meyer.

"I think everyone agrees the LAS is equitable, and it's doing what it was intended to do. It prevents waiting list deaths," said Dr. Golden.

The LAS system is reviewed periodically regarding organ allocation, survival data and other criteria to measure the success of the system.

Sources: UNOS and OPTN Web sites and physician interviews.

Lung Allocation Score for Lung Transplantation Leads to Increase in PF Transplants

Researchers at the University of Washington took a look at the Lung Allocation System to determine how pulmonary fibrosis patients, specifically, were faring since its inception. CPF Scientific Advisory Board member Ganesh Raghu, MD, was one of the study authors.

New findings published by University of Washington researchers indicate that the United Network for Organ Sharing (UNOS) Lung Allocation Score (LAS) system has led to an increased percentage of PF patients receiving transplants.

The research, Lung Allocation Score for Lung Transplantation: Impact on Disease Severity and Survival, was published online by the American College of Chest Physicians (ACCP), in April, 2008. The study was authored by Cynthia J. Gries, M.D., Michael S. Mulligan, M.D., Christopher H. Goss, M.D., Jeffrey D. Edelman, M.D., and Ganesh Raghu, M.D., from the University of Washington (Seattle, WA) and J. Randall Curtis, M.D. of Harborview Medical Center (Seattle, WA).

Researchers obtained data from UNOS on all lung transplant candidates listed and all patients undergoing transplantation in region six between May 4, 2003, and May 4, 2006. Each data set was divided into two cohorts: two years before LAS implementation, and one year after LAS implementation. Pre-LAS and post-LAS differences in patient characteristics were examined.

Researchers found that after LAS implementation, the distribution of diagnoses in patients undergoing transplantation significantly changed, while the distribution of diagnoses in candidates listed did not. More specifically, the study showed that a higher proportion patients received transplants (37.8%) were diagnosed with pulmonary fibrosis than prior to LAS Implementation (24.4%). Authors offered the possibility that more pulmonary fibrosis patients may be considered transplant candidates now that the LAS system is in place, because their disease severity may expedite the transplant procedure, whereas prior to the LAS system the patient may have been too ill to accrue time on a waiting list.

Characteristics of patients undergoing transplantation did not change, except that post-transplant hospital length of stay was shorter after LAS implementation. After controlling for age and diagnosis, neither waiting list nor transplant survival was significantly different. Researchers noted that larger and long-term survival studies are needed to determine if the LAS system improves overall allocation and survival for patients interested in lung transplantation.

Source: American College of Chest Physicians (Chest. 2007; 132:1954-1961); Content edited for space and clarity.

Study Suggests Rate of PF Progression Should be Considered as Trigger for Lung Transplant Referral Over Other Criteria

Lung transplantation is the only treatment modality that provides a survival advantage in pulmonary fibrosis (PF), but many patients deemed suitable will die awaiting lung transplantation. While donor organ shortage undoubtedly contributes to this, late referral to the transplant center may also play a role. A recent study investigated factors influencing the chance of patients with PF reaching lung transplantation.

Researchers at University of Newcastle (Newcastle upon Tyne, UK) conducted a single-center retrospective review of patient demographic data, assessment investigations and subsequent clinical outcomes was performed for patients with PF assessed for lung transplantation over a 5-year period. The results of the study appeared in a recent edition of *Thorax*, a peer-reviewed medical journal published by the British Thoracic Society.

Between March 1999 and March 2004, 129 patients with PF underwent formal transplant assessment. Sixty-nine were accepted and listed for lung transplantation. Of these, 17 were transplanted, 37 died while waiting, four were removed from the list and 11 were still waiting at the conclusion of the study. The median waiting time on the list for those transplanted was 103 days (range 6–904) compared with 125 days (range 2–547) for those who died while on the list ($p = 0.65$). There was no significant difference in age, spirometry, total lung capacity, gas transfer measures or six minute walk distance between those who died waiting and those transplanted. However, time from onset of symptoms to transplant assessment was significantly shorter in those who died on the waiting list (median 29 months (range 2–120)) than in those transplanted (median 46 months (range 6–204), $p = 0.037$).

The study authors concluded that patients with PF who died awaiting transplantation had similar disease severity at assessment as those who achieved transplantation. However, the interval between symptom onset and transplant referral was significantly shorter in those who died while on the waiting list, suggesting they had a more rapidly progressive disease. The rate of disease progression appears to be a more sensitive indicator for transplantation referral than any single physiological measure of disease severity and should act as an important trigger for early transplant referral.

Study Authors: Laura S Mackay, Rachel L Anderson, Gareth Parry, James Lordan, Paul A Corris, Andrew J Fisher of the Cardiopulmonary Transplant Unit, Freeman Hospital, Newcastle upon Tyne and Applied Immunobiology and Transplantation Research Group, University of Newcastle, Newcastle upon Tyne, UK

Former Marquette University Basketball Assistant Coach Transplanted, Transformed into Advocate

The CPF recently interviewed former Marquette University assistant basketball coach Trey Schwab about the transplant experience that not only saved his life, but changed his professional career.



Trey Schwab is an IPF transplant survivor who has dedicated his professional life to helping others awaiting lung transplantation.

When were you diagnosed? In the fall of 2001 at age 37.

What was your job before? Asst. basketball coach, Marquette University

When were you transplanted? Feb. 17, 2004

How long was your wait? I waited two and a half years. I was listed pretty much right away before the new lung allocation scoring system was in place. It was all based on time spent on the waiting list then. I automatically got six months credit on the list (PF patients were given credit due to disease severity.)

How has that new lung allocation system (LAS) changed things for PF patients? It is better now because those who are most sick get transplanted quicker. Previously, if you had a more aggressive doctor who put you on the list sooner, you got transplanted sooner. The primary factor now is how long you're expected to live without a transplant.

How long is the average wait for lungs on the transplant list today? The average waiting time ranges from 502 days to 659 days, depending on blood type. But the overall average wait time is under two years. Transplant centers are more aggressive in accepting lungs (from donors) and there are more organs available for transplantation.

How were your expectations for transplant different from the reality? They do a really good job at University of Wisconsin [where he was transplanted] of preparing you before hand. They show you and your family what to expect. They show you the room and the ventilator, they show you what you're going to look like so it is not as much of a shock when it happens. I had some severe complications, but overall it was pretty much what I thought it was going to be.

How did you get involved in the professional side of transplantation? I went back to Marquette for a year to continue coaching. That summer after the first season back things started falling into place. I wasn't out actively looking to change careers. It was a chance to help everyone who was still waiting. There was media attention to my job – even more so

after we made it to the Final Four. I wanted to help make the wait time shorter for the 80,000 people who were waiting on a list with me (98,000 now). I wanted to make sure others had the same opportunity I had. That's why I accepted this position at UW.

Do you miss sports? Sure. You don't do anything for 22-23 years and not miss it. I loved my job, but what I am doing now is important, too. I am still close enough that I can watch them play. The players still call and email me, so I'm still connected to them in a little way.

What can PF patients do once they are diagnosed if they want to be considered for transplant?

I always encourage anyone who has a serious disease like PF – you have a unique disease, you need a unique doctor. Unless you're at a large center that specializes in PF, you may not be getting the best possible care. Don't be afraid to change doctors or get another opinion. If you think you might be a candidate, call a transplant center and ask a coordinator. Most can at least give you guidance. Don't be shy. It is your life. If you're too sick, take along a friend who can ask the tough questions and be an advocate for you.

Is the new LAS system working better?

Of patients listed from 2004 to 2007, 60.7 percent of PF patients had been transplanted. Before the allocation rules changed, 51.6 percent of PF patients had been transplanted. This is proof the new system is working better for PF patients in particular because their LAS score is generally higher than other candidates.

Are there things a patient can be doing to increase their probability of being listed?

There are many things patients should be doing once they are diagnosed, and especially if transplant is a potential treatment option:

- **Ask your doctor** if transplant may be a treatment option if your disease progresses.
- **Call a transplant center** to seek information and determine if you could be eligible.
- **Participate in pulmonary rehabilitation classes;** the better shape you can keep yourself in the better. It will help you recover that much quicker.
- **Continue to be active.** I was working part-time the day I got transplanted and was going to rehab three days a week. I think that really helped me. Keep moving. Don't just sit and wait for the phone to ring – it will help both physically and mentally.
- **Be ready for a "dry run."** The lungs are only good for 3-4 hours. Often patients are called in for transplant procedure before the organ recovery takes place. Testing to make sure the organs are well-suited or healthy enough takes place and if the organ is found not suitable to use for transplant the recipient is sent home.
- **Get information.** Don't be afraid to talk to a couple of different coordinators and transplant centers. Most of the information is also available online.
- **Make an informed decision.** Different centers have different criteria. Don't take one center's "no" as an absolute final outcome. Ask another center. Circumstances can change. Keep seeking more information.

Research Shows Lung Transplant Allocation Scoring Process Achieving Objectives

A multi-center study was performed using data from five medical centers to review success of the Lung Allocation System. The study purpose to evaluate the impact of the lung allocation score on short-term outcomes after lung transplantation for all lung disease patients.

New research published in the Journal of Thoracic and Cardiovascular Surgery supports the fact that the lung allocation score is achieving its objectives.

The lung allocation scoring system restructured the distribution of scarce donor lungs for transplantation. The algorithm ranks waiting list patients according to medical urgency and expected benefit after transplantation.

The purpose of this study was to evaluate the impact of the lung allocation score on short-term outcomes after lung transplantation. A multicenter study was performed with data from five academic medical centers - University of Virginia Health System, Washington University School of Medicine, USC, the University of Wisconsin, and the

Mayo Clinic (MN). Results of patients undergoing transplantation on the basis of the lung allocation score (May 4, 2005 to May 3, 2006) were compared with those of patients receiving transplants the preceding year before the lung allocation score was implemented (May 4, 2004, to May 3, 2005).

The study authors concluded that the short-term outcomes of 341 patients (170 before the lung allocation score and 171 after) support the fact that the lung allocation score is achieving its objectives. Recipient diagnoses changed with an increase in PF and a decrease in emphysema and cystic fibrosis. The lung allocation score reduced waiting time and altered the distribution of lung diseases for which transplantation was done on the basis of medical necessity. After

transplantation, recipients have significantly higher rates of primary graft dysfunction and intensive care unit lengths of stay. However, hospital mortality and 1-year survival were not adversely affected.

Study authors were Benjamin D. Kozower, M.D., Bryan F. Meyers, M.D., Michael A. Smith, M.D., Nilto C. De Oliveira, M.D., Stephen D. Cassivi, M.D., Tracey J. Guthrie, RN, Honkung Wang, PhD, Beverly J. Ryan, ACNP, K. Robert Shen, M.D., Thomas M. Daniel, M.D., David R. Jones, M.D.

Source: J Thorac Cardiovasc Surg 2008;135:166-171; content edited for clarity and space.

Seminole, Florida Mayor Has New Lung, New Purpose

Mayor Jimmy Johnson returned home in the pouring rain just 12 days after undergoing lung transplant surgery. Friends and neighbors have rallied around him, preparing meals, making sure he takes his medicine and offering to do anything they can for him. "There's no place like home," Johnson said. Then, quoting a Kingston Trio song, itself a paraphrase of Robert Louis Stevenson, he added: "Home is the hunter, home from the hill ... It's wonderful."

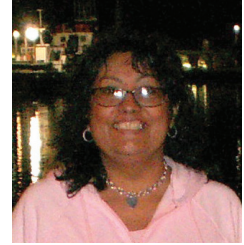
The outside world has called, too, asking the mayor to help push a bill before Congress that would increase funding for research into pulmonary fibrosis, Johnson's disease.

His plans don't stop at the doors to Seminole City Hall and the Chamber of Commerce, where he is executive director. He has been receiving cards and letters from across the world. Many of them refer to cystic fibrosis, an inherited disease that causes mucus to clog the lungs, or to the disease Johnson has, pulmonary fibrosis, a progressive scarring of the lungs. There is no cure for either disease.

Many people have never heard of pulmonary fibrosis. Johnson was one of those people until he was diagnosed with it in December.

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PF Transplant Patient Becomes Leader of Support Group, Advocate for Other Patients



Gloria Rodriguez was just 45-years-old when she heard the devastating diagnosis of PF. She was a mother and held a full-time job and simply didn't have time to be sick.

After three years, her disease began to worsen and she became completely oxygen dependent 24-hours a day. "I couldn't walk or talk," Rodriguez said, due to her extreme breathlessness. "I ended up in a wheelchair and my daughter had to bathe me. I became totally dependent on my family."

On May 5, 2005, Rodriguez was returning to her home in Los Angeles following a clinic appointment with her doctor at Stanford University Medical Center and another check up as she awaiting a lung transplant. The same day, she made her first visit to a support group which she says was the first time she'd met another PF patient and others on transplant lists.

The seven hour drive to Stanford was becoming more and more taxing for her. As she began the long trip home on May 5th, her mobile phone rang. The voice on the other end of the line told her to turn around and return the hospital. Her new lungs were waiting.

As is typical, there were pre-operation tests and a wait to determine if the new donor lungs were a good fit for her. They were. At 1:45 a.m. the next morning, Rodriguez went into the nine and a half hour surgery to save her life.

As Rodriguez recovered, spending three months at Stanford, she thought about the support group she'd attended the day she got the call for transplant. She knew the type of support she'd experienced that day would be helpful to her as she entered into a new life as an PF survivor and post-transplant patient. She found a support group not far from her home in

PF and transplant patient Gloria Rodriguez encourages other patients through an PF/transplant support group.

Los Angeles for transplant patients. She offered to take over the leadership of the group and her offer was accepted. "I knew what my family and I went through and I wanted to prepare those that were going to go through the same things I had," Rodriguez said.

Rodriguez started the support group with four people and has now grown the group to 35 people. She says each month she gets new referrals. "It has been a life changing experience for me," she said.

If you live in the Los Angeles area and would like to join Rodriguez' PF and transplant support group, contact her at: Gloria Rodriguez (562) 941-2368 or via email at Glorry2God@aol.com.

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Raising awareness is one of the goals of the California-based Coalition for Pulmonary Fibrosis, which contacted Johnson after reading a news article about him.

"He really is an inspiration. He's also a minority. There are very, very few of (our patients who) survive," said Teresa Geiger, the coalition's vice president of outreach and patient advocacy. Geiger's father died of the disease. "This is a diagnosis that is worse than lung cancer," Geiger said. "It's a tough disease. It's a tough diagnosis."

Geiger has asked Johnson to help her advocate a bill that's being sponsored in Congress by U.S. Rep. Brian Baird, D-Wash. Baird had a relative with the disease.

The bill, if passed, would do four things: create a pulmonary fibrosis registry; set up a national pulmonary fibrosis advisory board to make recommendations to the Department of Health and Human Services; expand research of the disease at the National Institutes of Health; and require the Centers for Disease Control to prepare a national action plan.

Source: St. Petersburg Times; Content edited for space.

Loyola Transplant Surgeon Pioneers New Way to Increase Organ Supply

Transplant surgeons such as Dr. Robert Love of Loyola University Hospital are pioneering a new way to increase the supply of organs from deceased donors.

Today, most organs are obtained from donors on life support who have been declared brain dead. The organs are in good condition because the heart is still beating. But more surgeons are beginning to use organs from patients who have been declared dead after their hearts have stopped beating. Last year, there were 793 donations after cardiac death in the United States. That's up from 117 cases in 2000.

Kidneys and livers are the most commonly used organs from cardiac-death donors, but surgeons also are using lungs and pancreases. Love has done about 30 lung transplants from cardiac-death donors. Nearly 100,000 people are waiting for organ transplants in the U.S. Some people wait for years. And many die while still on the list.

"We must do everything we can to encourage people to become organ donors," Love said. "And we also have to make sure that every usable organ is used." Love is a professor of thoracic and cardiovascular surgery at Loyola University Chicago Stritch School of Medicine.

Obtaining organs from cardiac-death donors can be technically challenging. Organs begin to deteriorate as soon as the heart stops beating. Surgeons generally have only a 30-minute to 60-minute window to remove organs. Five minutes after the patient's heart stops beating, the body is taken to an operating room, where the organs are recovered.

Donation after cardiac death leaves the family little time to say goodbye. "But families are usually very understanding about this," said Joyce Maly, Loyola's in-house coordinator for organ and tissue donation. Maly's position is a partnership between Loyola and Gift of Hope Organ and Tissue Donor Network.

Source - Newswise; Content Edited for Space.

"You are providing an unbelievable helpful combination of educational materials and support to patients – keep up the good work"

– ALA of New York

Transplant patient Mike Henderson Shares Experience and New Role as Advocate and Fundraiser

When were you diagnosed with PF and what was your experience early on? I was diagnosed in June of 2004 and initially treated by a pulmonary specialist in Portland, Oregon. After my diagnosis I began to research the internet and learned that several clinical trials were underway and that Dr. Ganesh Raghu at the University of Washington Medical Center was a world renowned expert in PF. I also learned that I should immediately pursue placement on the lung transplant list. I was referred to Dr. Raghu, as well as the Lung Transplant Program at Oregon Health Sciences University.

Were you placed on the transplant list right away? No. I met with Dr. Raghu in December of 2004 and considered the courses of action available to me. I was pleased to learn that the waiting time for a lung transplant at the University of Washington was three to six months, while waits were two to three years at some institutions around the country. I asked Dr. Raghu to put me on the list and he said it was not time yet. It was a bit traumatic because the first test was an angiogram and it identified a coronary artery blockage, so a stent was placed. Unfortunately this also meant a six month delay. So, I was officially turned down by the committee, but encouraged to complete the rest of the tests and to have another angiogram in six months. Several cardiologists determined that my coronary artery disease was stable and I was approved for transplant January 4, 2006. I did receive two new lungs on

March 19, 2006, about two and a half months after being listed.

What was it like going through the process of being evaluated for transplant and being listed?

Other than having every part of my body, mind and family history and environment probed, it was hardly invasive at all (just kidding). As far as my family, we tried to make it an adventure. We moved from Portland to downtown Seattle. We enjoyed re-connecting with family and friends, trying new restaurants and walking as far as I was able. It was a great adventure, interrupted by two “false alarms”. We found out how unprepared we were the first time, running around like chickens trying to figure out what to take with us. We put the kids on alert the first time, but didn’t call until we knew it was real the next two times.

How important is a support system (family, friends) when you’re going through transplant?

It was unbelievably helpful. All my kids were able to be there to see me off to the operating room and all still there when I came back to life the next day. My family, my wife’s family and several close friends were in constant contact and cheering me on. The most important and most helpful was my wife Donna. She never wanted to be a nurse, but I would never have made it through without her.

Describe the transplant experience, if you can. What was it like going through it, physically and emotionally?

I had thought a lot about whether it made sense for me to pursue a transplant and it boiled down to one issue, quality of life. I was comfortable with death, since I had lived a very blessed life, but

felt that if I could have a few years of quality of life, it was worth the risk. So, when the day came, I was very comfortable with whatever the outcome.

What would you say to other patients who are potential transplant patients about the risks involved?

It is definitely a high risk, high reward situation. Reflecting back, the surgery itself seems less risky than the compatibility of the matching lungs, the impact on my body overall, and the adjustments to the drug routine. If you have problems coming in, you will exacerbate many of them coming out of surgery. It really tests the strength of your relationships. It’s not for everyone, but I would do it again.

How did your experience as a patient move you toward becoming a patient advocate?

After recovering from the transplant, I thought about what I went through when I was first diagnosed. I had no idea what PF was and no idea what options were available to me. I attended a Pulmonary Fibrosis Support Group meeting and met others with the disease. The support group was tremendously helpful to Donna and me, so we decided to do our best to help others deal with this devastating diagnosis. We took what we learned and helped others get a support group launched in Portland.

Why did you decide to partner with the CPF for an educational event?

I learned that Dr. Raghu and the CPF had sponsored an PF seminar in Seattle several years earlier and that the group was anxious to have another one. So, I committed to lead that effort. Fortunately, and unfortunately, my efforts were

interrupted by a lung transplant. When I had my strength back, I learned that the CPF had helped sponsor a number of similar events around the country. I felt their event experience and promotional capabilities would be very helpful.

How did you raise funds for a patient event in Seattle?

It was far easier than I expected. I composed an email asking for support and provided a link to my own fundraising page. I sent it to friends and family and raised nearly \$18,000 in six weeks.

Are there tips you have for others who’d like to fundraise for the CPF?

The Internet is a wonderful tool. Whether you do it the way I did, by just asking for a donation, or by creating an event where people can pledge money per mile, it is a very efficient way to raise money for a great cause. You will be amazed how generous people can be and surprised at who really steps up to support you.

Are there other things you’d like to do with the CPF to help raise awareness and funding for PF?

Our support group in Seattle is brainstorming that very issue at our next meeting. Stay tuned!

How would you encourage others to become involved?

The most important thing is to spread the word about PF. I think the CPF does a great job nationally and we need to support them and also get them involved on a local level. We need to raise awareness first and money for research and treatment will follow.

Transplanted Daughter Wishes Mother Could Have Same Gift of Life

A Colorado woman breathes easier in the mile high city of Denver, just months after receiving a life-saving lung transplant from an incurable lung disease called pulmonary fibrosis (PF). She only wishes her mother, who suffers from the same disease, could receive the same gift of life.

Terry DeLeon, 56, was diagnosed with PF just a month after her mother, Laura Sides' diagnosis in late 2005. She'd been helping her mother deal with her illness and news that the disease was terminal when she started to notice that she, herself, was experiencing the same symptoms – a dry cough and shortness of breath.

A high resolution computer tomography (HRCT) scan revealed what she feared. She, too, suffered from pulmonary fibrosis. It is believed that 15 percent of PF cases are genetic in origin. Sides lost her sister, a half-sister and a half-brother to PF.

DeLeon quickly deteriorated, much faster than her mother's condition. Within months of her diagnosis, she was completely oxygen dependent. She received her lung transplant on August 4, 2007 after being listed for transplant in May 2007.

DeLeon says she feels a little guilty about being the one who was transplanted. "My mom doesn't have the opportunity to get a lung because of her age and other health conditions and that makes me feel pretty bad. I don't know how else to describe it. I wish I could do the same for her that was done for me. It is so humbling."

But Sides believes her daughter should have been the one with the second chance at life. "To see her suffer was the worse part for me. She suffered so much more than I have. When her breathing went, it was hard for me to see that because I know what it is like and how hard it is," she said. "My suffering is nothing compared to hers. It is more important for her to get what she needs. I have put up with knowing what this disease does and knowing that I am going to die anyway from it. But she has a chance. It didn't bother me nearly as much as it bothered her."

DeLeon's condition has improved dramatically since her transplant, though it has not been without minor setbacks. "It took me about six months before I really felt like getting up and getting around and doing normal lifestyle activities. I gradually got to where I could stand



IPF transplant patient Terri DeLeon with her mother, Laura Sides, also a PF patient.

long enough to do the dishes. I purposely didn't take it too fast. I am the type of person that will over do it if I am not careful," she said.

She's happy, she says, that she can again do some of the things that PF took away from her. "One day, I had been listening to my mp3 player and I could sing along with it. I just bawled. That was something I thought I had lost."

"My mom doesn't have the opportunity to get a lung because of her age and other health conditions....I wish I could do the same for her that was done for me."

*– Terri DeLeon,
PF transplant patient*

Currently, the National Institutes of Health is sponsoring a multi-center clinical trial investigating the use of Viagra (sildenafil) in pulmonary fibrosis.

To find out more about the study and study locations nationwide, please visit www.ipfnet.org or call (800) 447-3638.

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Lung Transplant Resources

Visit the CPF Web site for a listing of transplant centers with PF specialties:

www.coalitionforpf.org/AboutUs/resources_websites.asp

General transplant information:

www.unos.org

www.transweb.org

Information regarding organ and tissue donation:

www.sos.state.il.us/depts/drivers/programs/donor/donor.html

Other resources:

National Transplant Society: www.organdonor.org

TransWeb: www.transweb.org

TRIO: www.trioweb.org



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