

Chapter Six:

Lung Transplantation



- 6.1 Lung Transplantation
- 6.2 Funding Your Transplant
- 6.3 Promoting Organ Donation

6.1 Lung Transplantation

Although there is now an FDA approved treatment for LAM, there is still no cure. Many women with LAM remain stable for years but most learn sooner or later that LAM is a progressive disease. Whether your progress is fast and aggressive or slow and gradual, there may be a chance you'll reach a point in time when you'll consider having a lung transplant. A lung transplant is an amazing surgery and it requires many life changes that demand careful thought and lots of self-education.

When Is It Time to Consider Lung Transplantation?

If you reach the point where your disease has progressed and you're struggling with day-to-day chores, you'll probably find yourself thinking about undergoing a lung transplant. You might find that you need additional supplemental oxygen to perform even simple tasks, or you might notice that you have a decreased capacity for exercise and/or that you tend to get fatigued far more easily. Keep in mind that these changes can occur even without big changes in your pulmonary function tests. Eventually, you or your doctor will know that it is time for you to be evaluated for transplant.

Your doctors will probably bring up the topic of transplant when they feel that your LAM has reached a certain level of severity. There are some concrete data to watch and your doctor should keep an eye on for you. Some of the main indicators are your PFT results, especially your FEV1 number. Many pulmonary specialists feel that evaluation for lung transplant is appropriate when your FEV1 drops below 40% of the predicted normal score. Other important numbers are your DLCO, an indicator of how well your body gets oxygen from your lungs to your blood, and PaO₂, the number, resulting from an arterial blood gas test, that indicates how much oxygen is really in your blood. When either your DLCO or your PaO₂ falls below the sixty-percent range, you probably need to start paying more attention to the numbers. Finally, your body's response to supplemental oxygen (O₂) is another important factor and is related to the DLCO and the PaO₂.

But the answer to the question "When is it time to get a transplant?" rests largely with you and has no hard and fast facts to back it up. For some LAM patients, the answer is "Never! I'll take my chances and live my life without the added risks inherent in transplantation." This is a valid answer. Each woman has the right to make this decision.* Your own self-awareness is essential in choosing if or when to have a transplant because you are the best judge of your quality of life. You are the one who lugs the O₂ tanks, shuns the stairs and takes breathing breaks from simple everyday tasks. When you feel the risks of a major surgery and the additional complications that result after transplantation would be better than your current situation with decreased lung function, then it might be time to say, "Okay, let's talk about transplantation." That will be your first step on an incredible journey.

* If you think you don't ever want to undergo a transplant, you should still get evaluated and get listed just in case you change your mind should you suddenly become much sicker. You can always say no to a transplant later.

Choosing a Hospital

Not all hospitals perform lung transplants. There are standard criteria for a hospital to be qualified for transplants in general and then for lung transplantation specifically.** Your pulmonologist's opinion on transplant centers is an important starting point. Geographical proximity (the support of your family and friends who can be close by is very important), the length of the waiting list and the hospital's record of success with lung transplantation are other factors that you should consider. Your health insurance has a big say in this decision, too.

Evaluation: Will I Measure Up?

Your doctor will send a referral letter and your medical records to the transplant center you have chosen. If the experts there feel that you are a potential transplant patient, one of them (probably the one you'll come to know as your transplant coordinator) will call you to schedule your evaluation procedure. You'll go to the hospital, probably as an outpatient, undergo many tests, and then have several important meetings to review the test results.

As crazy as it sounds, your evaluation largely will be to determine not if you're *sick* enough to need new lungs but if you're *healthy* enough to receive them! Lung transplantation is a very complex surgery and life after transplant brings many additional health concerns and hazards. Most of the medical tests in your evaluation will focus on your overall health, especially the condition of your heart and kidneys. These organs, more than all the others, must be functioning well. A heart catheterization will probably be performed during the transplant workup to ensure that your heart is up to the stress of the surgery. Your kidney function will be evaluated to make sure your kidneys are up to processing all the immune-suppressant drugs after transplant.

The evaluation will also include an electrocardiogram, an echocardiogram, lab tests (blood and urine workups), PFTs, and CT scans. You may also be required to have a Pap smear, a mammogram, or other cancer-screening tests. Additionally, you'll be checked for infections like HIV and hepatitis. Other tests that may be added include a stool test, a TB test, and a dental evaluation. You'll also have your bone density checked to see if you suffer from osteoporosis. If you do—and many LAM patients do—you'll need to begin treatment for this before transplant. Because post-transplant drugs can cause loss of bone density, making sure that your bones are as strong as possible before transplant is a necessity.

Although many transplant hospitals will require you to have all these tests done at that particular center, some will accept certain test results from your personal hospital or from the NIH.

A transplant evaluation does not just include medical tests. You'll meet with a financial counselor to discuss whether your insurance will cover the cost of your surgery,* your follow-up care, and your post-transplant medications. You'll also meet with a social worker to talk about the emotional stresses of the process that you and your family will invariably confront and to make sure that you have the necessary emotional and physical support. Your social worker may suggest an anti-depressant or anti-anxiety medication for you or for your family members to alleviate some of the normal stress associated with a transplant. Because mental stress

** Contact the United Network for Organ Sharing (UNOS) office if you want a complete list of hospitals that are qualified to perform lung transplantation (1-888-TXINFO1 or <http://www.unos.org>).

* See the chapter on Funding Your Transplant for help in financial concerns.

translates into physical stress, don't hesitate to use these medications, and possibly sleeping pills, to get you through this time period.

These discussions with the social worker are also necessary to insure that you are someone worthy of new lungs, that you'll follow your doctors' instructions and treat your new lungs as the very rare gift that they are. A dietician and an exercise therapist round out the evaluation process to make sure you will be in tip-top shape when your time comes for transplant. Transplant centers typically want you at a normal weight—neither obese nor malnourished. They also prefer you to be ambulatory, since being able to walk allows you to recuperate from the surgery more easily.

A final note about transplant readiness: **NO** transplant center will transplant donor lungs into the body of a smoker. If you smoke, you must quit. Many centers require you to be a nonsmoker for at least six months before they will consider accepting you. Other hindrances to receiving new lungs are excessive alcohol consumption and the use of narcotic medications. All three of these issues must be addressed before you are considered for a transplant.

You'll have many chances to ask questions, and you'll learn both bad news and good news. The bad news is that a lung transplant is not an automatic ticket to "Health Heaven." With a lung transplant, you trade one set of problems for another. After your transplant, your immune system must be suppressed to help your body accept its new lung(s). You'll take many medications that will place you at higher risk for some other medical conditions such as kidney disease and diabetes. The list of possible complications can seem overwhelming, but you must face them and be willing to accept the risks (or at least be ready to start getting used to the thought of them!). In exchange, the good news is that you'll be able to BREATHE! You'll experience deep, trouble-free breathing. You'll gain renewed mobility and energy and develop greater stamina. Yes, the cost of a lung transplant is high, both in dollars and in physical risks, but many LAM patients who have had a lung transplant will tell you that their new lives have been worth every bit of the price.

Finally, at the end of the evaluation process, which can last from a couple of days to a week, you will have an assessment meeting with your transplant coordinator and other members of the staff. They'll go over their findings and tell you if you've been accepted as a transplant patient. This team will discuss the probable length of your wait and tell you what you can do in the meantime to prepare yourself for this medical miracle called lung transplantation.

Will I Get a Single- or a Double-Lung Transplant?

This seems to be the million-dollar question. No one can say for certain whether you'll get one lung (a unilateral transplant) or two (a bilateral transplant) until you get the call, but certain factors govern how many lungs you get. Some transplant centers believe that all LAM patients should ideally have double-lung transplants, but that doesn't mean you will be guaranteed two lungs. It's always a question of availability. If you're healthy enough to wait for two lungs, you may be able to hold out for several years waiting for just the right pair. However, you may be so sick that you need to get the first available transplant just to live, even if that means you'll only receive one lung.

In the past, having had pleurodesis on a lung meant that you could only get a single lung. That's no longer true. While there is a greater risk of bleeding and the procedure may take a bit longer if your lung has been pleurodesed, it does not typically lead to a longer hospital stay

after transplantation. Moral of the story – a previous pleurodesis will not prevent you from having a double lung transplant.

Living-Lobar Transplants

If LAM is progressing rapidly and you and your doctors don't feel you'll survive the wait for a cadaveric transplant, you have one other option: a living-lobar transplant. In this type of transplant, a lobe is removed from each of two family members (or suitable donors) and transplanted into the lung recipient. This method of transplant is possible because each person has a total of five lobes: three in the right lung and two in the left. Healthy individuals usually have no decrease in breathing ability after giving up one lobe of their lungs.

This type of transplant is rarely the first option because of the risks not just to the recipient but also to the donors and because of the possible emotional repercussions in the family should problems arise with those giving or receiving lobes. In the past, this procedure has generally been reserved for children (the lobes, being only sections of lungs, are often too small for adults). But progress is being made all the time, and living-lobar transplants for adults are becoming more successful. However, finding two family members (or suitable donors) who are both willing to donate a lobe of their lungs *and* who are compatible with the recipient is a huge challenge.

However, there are some distinct advantages to this method. First, your waiting time for lungs may be shortened considerably. Second, because these lobes often come from family members, your chances for rejection may be significantly reduced. And third, the surgery will be planned in advance so that the donor lobes can be transplanted far more quickly than a standard transplant, insuring a healthier condition for the lobes.

If you're becoming so sick that you're worried about not receiving a transplant quickly enough, ask your doctors at your transplant center about this option.

Waiting for the Call

After your evaluation is finished and you have returned home, you will resume the life you had been living before the evaluation. To a great extent, nothing will be changed about what you do and how you live. The only difference, for many months, will be that you know that there is something special ahead for you in your future. You'll need to watch your weight and exercise faithfully. This is also a time to continue your social contacts and to do as many of the activities that you always wanted to do and still can do. You also need to take this time to prepare for any possible outcome. Get a living will, a power of attorney and get your legal affairs in order, just in case.

You will also want to contact The LAM Foundation to discuss how you can donate your old lungs to research. Live LAM cells are the best material available for our researchers to use when looking for how the lungs will react to different treatments. The process for donating your tissue is simple and will include contacting NDRI, the organization that manages the procurement and distribution of the tissue, filling out some paperwork and calling them when you receive the call for transplant.

You'll hear from your transplant coordinator from time to time, and the transplant team will ask you to return for regular check-up tests. Some centers may permit you to be checked by your local physician if he or she is willing to forward the assessments of your status to the transplant center. As your waiting time accrues and as the time for transplant grows closer, the

transplant center may require you to move to its immediate vicinity if you live too far away.* The social worker there can help you find housing near the hospital. Other transplant centers simply require that you insure your prompt arrival at their hospital as soon as you get your call.

When you know your call is coming soon, make some specific plans that can simplify your life when the call really comes. Make sure important phone numbers are programmed into your cell phone. When you get your call, no matter where you are, you'll be ready to begin making crucial calls. Some of the people you may want to contact are your family and friends, your doctor, your transplant coordinator and your pilot if you'll be using a service like Angel Flight* or a corporate jet. Make sure each member of your family and all of your friends have additional phone numbers to contact others. *And don't forget to notify NDRI* so they can begin to make arrangements to collect your old lungs. Waiting for the call is stressful in and of itself. You might have one or more dry runs—when you rush off to the hospital only to find that the donated lung (or lungs) isn't a good match or that it's damaged in some way. Just take a deep breath (okay, take as deep a breath as you can) and go back to as normal an existence as possible.

This waiting period is also the time to make childcare arrangements if your children are small, find someone to care for your pets and give one of your neighbors a key to your house to water your plants and take in mail and newspapers. Will you need someone to mow the lawn or shovel snow? Make arrangements for those things too.

Pack a survival bag for the people who will be going with you and who will be waiting at the transplant center. Even though you'll be undergoing major physical stress to your body, you'll be unconscious. Your supporters, however, will be awake and very anxious while you're in surgery, which can take 6 - 12 hours or more. Your survival bag might include snacks—munchies, gum and hard candy – and consider including a few rolls of coins for vending machines. Make sure you pack all the names, phone numbers, and e-mail addresses of people who will want to know your condition. Magazines with short articles or games and puzzles will help those waiting to pass the time. Even a deck of cards or board games like Scrabble or Monopoly will help keep your family members occupied and distracted as they wait. Make sure to include a pad of paper and some pens and pencils. Your family will want to take notes on your progress so they don't forget to tell any important bits of news to everybody they call.

Don't forget your needs when you're getting ready. Although your transplant coordinator may tell you to bring nothing at all with you when you first come to the hospital, one of your caretakers will need to bring some things to you after your surgery. Keep a suitcase packed just like you did when you were pregnant and waiting for labor to begin. Pack pajamas with a button front, or just bottoms (you don't need a draft behind you) to wear with the hospital gowns. Keep in mind that the hospital gowns with snaps work better than pajama tops when you still have an IV in.

Remember to take whatever you need to make you feel as normal as possible when you're recovering after the surgery. Do you need a special tea to soothe your nerves? What about a favorite scented body lotion to mask that hospital smell? A pretty lip-gloss might make the difference between a good day and a bad one. Pack what you need to use after transplant to

* Generally, you must live no further than a four-hour drive away, and preferably closer.

* Angel Flight is just one of several nonprofit services that carry individuals who are sick or injured to hospitals for a minimal fee or free of charge.

make you feel good, pretty and perky. You'll have earned these little rewards, and these small preparations can help you feel better after your surgery.

The Big Day

When you finally get the call, you'll need to get to the hospital as soon as possible, usually within a few hours. Once there, you'll get a physical. If you have a cold or any sort of infection, you'll not receive the transplant. If you pass the physical, you'll have IVs inserted, and you'll be prepped for surgery. And, don't be surprised if it's a case of "hurry up and wait." Even after you're prepped, you might have to wait up to six hours for an operating room or for the lungs to be delivered. You might have another dry run. But keep waiting. The time will eventually come.

After Transplant

After the surgery, you'll wake up in the Intensive Care Unit (ICU). You'll have an endotracheal tube in place, some chest tubes (usually two for a single-lung and four for a double-lung transplant), and various IV lines and other monitors. You'll feel as if you were hit by a truck, but you'll be able to breathe, and that lovely sensation will do wonders for your mood. Part of your discomfort will be caused by the medications pumped into you during the surgery, and those same meds might make you feel a bit delirious or even confused for up to a week or more. These problems will quickly begin to subside. Despite the pain, try to remember that the most severe discomfort will probably decrease in 24-48 hours. The endotracheal tube will generally come out quickly, but your chest tubes will remain in for a while to aid drainage. If you're doing well, you may be moved to a ward in 48-72 hours.

Once in the ward, you'll be expected to start moving and using your new lungs. Just when you want to rest for a long time, the staff will get you sitting up and walking around. Before you know it, someone will make you walk on a treadmill or work out on a stationary bike. You'll have to work to get your new lung(s) functioning at top capacity.

You'll also be given a variety of new medications, some of which may be started just before the transplant, to stop rejection and prevent infection. You'll get used to them in time, and as your body adjusts, your doses will be increased or decreased. Don't be discouraged if it takes some trial and error to find the dose or the correct combination of drugs that works for you. Eventually your doctors will adjust your medications to the point that you'll feel like yourself again.

You might find that your body tries to reject your new lungs. When this occurs shortly after transplant, it's called acute rejection and it means that your body is mounting an attack on the new organ at the cellular level. Should you worry? Yes a bit, but acute rejection can be treated very successfully with medications. It's all part of the process of your body accepting your new lung(s). Later, you might suffer from chronic rejection. This type of rejection is your body's defenses fighting in the airways. This, too, is treatable with drugs.

In the first three months, you'll find that you have far too many drugs to deal with. Learning what drug to take when will come with practice, but you'll also need to learn warning signs of potential problems so you can ask your doctor whether adjustments in your medications are necessary. During these early months, you might continue to suffer from nausea, confusion and swelling in your legs. You'll also have routine bronchoscopies performed at the transplant center. A bronchoscopy is the only way your doctor can get direct access to your lungs, access

that makes it possible to monitor your transplant for signs of rejection and infection. You'll probably have two to six bronchoscopies in your first year after transplant; they're that important.

You'll probably also be given a small home-use spirometer to check and chart your FEV1, another tool to monitor your body for rejection. While you're charting your FEV1, write down how you feel that day and whether you have a fever or any respiratory symptoms. After your transplant you can't really wait until the next day as you used to. Once you've had a transplant, these little problems become big ones very quickly.

In three to six months after transplant, you should be driving again and you'll probably resume many of your normal household activities. Don't overdo it. Rest when you need to and allow your lungs to strengthen slowly. Don't be upset if your household feels topsy-turvy. Your family has been making do without you for some time, and it will take all of you to settle into a comfortable routine again. Talk out problems as soon as they come up, and seek professional counseling if they continue.

Life after Transplant

You've done it. Congratulations! Go home and take good care of your new gift. Take your medications, exercise, eat well and enjoy your new lease on life.

Follow your doctors' orders on how fast to re-enter life. Take it slowly so that you don't harm your new lung(s). After transplant, you'll be very susceptible to infections so you may have to avoid crowds for a while. You'll also need plenty of time for your body to heal fully from the surgery.

It took you many years to get as sick as you were, and it may take a few years to regain your former health. Each woman is different, and each transplant is unique. You might have FEV1s in the 60s or 70s for a few years as your lungs build up their stamina, or you might feel terrific in a month and have an FEV1 of 100%. Don't be discouraged if your progress is slow. You *will* get better. You may never be where you were before you had LAM, but you'll make a miraculous recovery.

Lung transplantation is, indeed, an amazing process. As waiting lists grow longer, many LAM patients are being encouraged to start the process fairly early in their lives with LAM. You must always remember, however, that transplantation isn't a cure. Consider it an opportunity for a better life, and prepare for it carefully.

Recommended Reading:

The Lung Transplantation Handbook: A Guide for Patients

Author Karen A. Couture, a LAM patient and double-lung recipient, has written a lifesaving guide for transplant patients. The "how-to" handbook introduces its readers to the complex process of getting on the transplant waiting list and how the list works; preparing for surgery and the surgery itself; financing the operation and the long recovery afterward; and the complications of rejection, infections and the medications. Scattered throughout the book are short, but inspirational stories and quotes from patients who have gone through this before. The book also provides a wealth of information in an extensive resource section; a glossary; and an appendix of all lung transplant centers in the United States. Order the book online at

www.trafford.com (search for *The Lung Transplantation Handbook: A Guide for Patients*) or call 888-232-4444.

Helpful Websites

Medline Plus, a service of the U.S. National Library of Medicine and National Institutes of Health
<http://www.nlm.nih.gov/medlineplus/lungtransplantation.html>

Duke Transplant Center

<https://www.dukehealth.org/treatments/transplant-program/lung-transplant>Cleveland Clinic

Lung Transplant Program

<http://my.clevelandclinic.org/services/transplant-center/transplant-programs/lung-transplant-program>

6.2 Funding Your Transplant

Your insurance may cover the entire transplant and all of your medications or it may not. But even if it does cover most items, you'll still have lots of additional financial concerns such as time off work for you or your caregiver(s), lodging near the transplant center before and after transplant, childcare and/or pet boarding, extra mileage for all the trips to the hospital and all the fast food eaten in the hospital by your loved ones. If you prepare well beforehand and set up all of your funding before your surgery, after transplant you'll only have to worry about getting better.

There are several nonprofit agencies that will help you fund your transplant. If you need financial help, search the Web for a suitable agency. Before formally working with any group, research and question each organization for their commitment to helping you.

Transplant Living

A good place to start your search is The United Network for Organ Sharing (UNOS). This non-profit organization sponsors a website called Transplant Living. The site has a wealth of information on transplants in general and a thorough section on financing a transplant too. Go to the Transplant Living website (<http://www.transplantliving.org>) and pull down the menu *Before the Transplant*. Then click on *Financing a Transplant*, and you're on your way. The site discusses insurance, Medicare, fundraising, prescription drug assistance and many more ways to pay for your expenses.

HelpHopeLive

HelpHopeLive is a nonprofit organization that can help you pay for any transplant-related expenses such as transplant medicines, relocation expenses and mileage to and from all those pre-surgery visits to your transplant center. It can even help you pay for the transplant itself!

Here are the basics of how the organization works. You contact the staff at the HelpHopeLive and they set up a tax-deductible account in your name for the donations you'll receive. Then they'll help you generate ideas to raise money by providing information on how to hold various fundraisers from online fundraising pages to spaghetti dinners to auctions to galas. They'll even help with promotional materials.

When you raise any money, you put it into this special account and because the organization is a 501(c)(3) nonprofit, all the donations are tax-deductible.

You personally receive several benefits. First, the money goes to the nonprofit fund, so you are not taxed on it. Second, when you incur expenses, you simply send the bills to the HelpHopeLive and they write out the checks and mail them. What's in it for the HelpHopeLive? They want you to spread the word about organ donation. It's a win-win situation for both you and HelpHopeLive. To learn more, give them a call at 1-800-642-8399 or check out their website at <https://helphopelive.org> today.

**Don't wait until you're too sick to help fund your transplant.
Check out funding options as early as you can.**

6.3 Promoting Organ Donation

One complicating factor in the timing of your transplant is the scarcity of donated lungs. Once you and your doctors decide that you're ready to proceed with an evaluation and listing, you'll find that hundreds, maybe thousands, of people are ahead of you on a waiting list for healthy lungs.

The list is specific to the hospital you choose, but it's coordinated with all of the waiting lists at all of the transplant centers across the US. The United Network for Organ Sharing (UNOS) works under contract with the federal government and oversees the procurement and disbursement of donated solid organs nationwide. Once you have been evaluated and approved, your name will go into the UNOS databank as well as onto the waiting list at your chosen hospital.

Of all the solid organs that can be successfully transplanted, lungs are the most fragile. They are easily damaged in car accidents and other fatal incidents that most often bring donors' organs into transplant hospitals. Lungs must be matched between donor and recipient not only by blood type but also by size. You may find yourself at the top of your list only to wait several more months until the right lungs come along for you. Because this waiting period can be so precarious, it's essential that every LAM patient and all families and friends of LAM patients *become active in the promotion of organ donation*.

The LAM Foundation actively promotes organ donation because it understands the value of these precious gifts. Becoming an organ donor is easy and one of the most important and generous things you and your loved ones can do. Contact your local organ-procurement office (Google "Organ and Tissue Banks"). Second Wind, the national support organization for lung transplantation (<http://www.2ndwind.org>), is another good source for materials. Gather some information and some free brochures then get out there and get noisy! Promoting organ-donation awareness can mean the difference between life and death for any of the women who live with LAM. But promoting organ donation benefits all people facing serious medical challenges, not just LAM patients. Organ donation is an opportunity to change untimely loss to new life. One organ donor can touch the lives of up to fifty people who are waiting for "the gift of life." Consider becoming a donor yourself and encourage your family members to do the same.

If you decide to become a donor, you *must* let your family know your wishes. If you're on life support, the attending physicians will ask your family's permission to donate your organs—

even if the physicians have a card or your driver's license that you have signed stating your wishes. If your family members don't know that you want to be an organ donor and they say "no," your generous intention will be lost.

Remember the two steps: one, make your decision and two, tell your family.