

# THE LAM HANDBOOK

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A REFERENCE GUIDE FOR WOMEN WITH LAM







## **Dedication**

To all of the women with LAM,  
especially those who took the first step for all of us  
and who continue to lead us along the way.

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### **Medical Disclaimer**

This handbook is for educational purposes only. The information contained in this handbook should not be used to diagnose or treat a health problem or disease. This handbook should not be considered as offering medical advice and is not a substitute for consulting a licensed medical professional. You should rely on your physician's advice for the treatment of LAM or any of the related problems discussed in this handbook.

While we have endeavored to make sure that the information contained in this handbook is accurate, we cannot guarantee the accuracy of such information and it is provided without warranty or guarantee of any kind.



## Introduction

Lymphangioliomyomatosis. Bet you never thought that word would roll off your tongue. But now you're reading this handbook and there is both bad news and good news for you. The bad news is that you, or someone you love, has been diagnosed with LAM. The good news is that you are not alone. As you learn about LAM, you'll become acquainted with some of the kindest, most courageous and most caring women on earth. They'll become your fellow travelers on this most remarkable journey.

Although certainly not comprehensive, this handbook is meant to help you with various terms associated with LAM. There is a whole new vocabulary for you to learn. When you first hear some of these medical terms, you probably won't remember them but in time they will become quite familiar. We hope our explanations help you. Check the glossary for terms you don't understand or terms that aren't explained immediately in the text. Use the alphabet soup section for all of those annoying abbreviations and acronyms you can't remember. And if you don't find a word or an abbreviation that you know about, send a note to The LAM Foundation and we'll consider it for the next edition.

This handbook will answer many of your questions and offer solutions to some problems you may encounter. It provides tips on where you can learn more and enough information to know what questions to ask. You don't need to read this whole book from cover to cover right away. Just use it as a reference book when you have questions and concerns. We've tried to give a good overview of LAM and to make this a user-friendly manual but we've avoided giving advice on treatments and medications because, as research continues, these recommendations continue to change. And besides, no two women with LAM are exactly alike. We've tried to anticipate and answer your questions and we've attempted to prepare you for whatever lies ahead for you. Naturally, there are also questions to which there are no answers.

For those who are just beginning the journey, we welcome you to this community. As you become more familiar with LAM, you'll become guides for others. This is a small group of exceptional women who need and value each other's support. Though you may be a novice now, you'll soon be well versed in LAM. Don't hesitate to ask questions. For those who have coped with LAM for years now, this is *your* handbook. Many of you have unknowingly written or contributed to vast sections of it. Although we've derived much of the information from the scientific and medical communities, a great deal of our wisdom has come from you. You are the pioneers of LAM and LAM research and you've generously shared your experiences and knowledge with your fellow travelers. We've compiled bits and pieces from many, if not all, of you – at the NIH, at LAMposium, through letters, phone calls and through social media. Many of your little nugget and words of wisdom are lovingly presented here. We give all of you credit and thank you for your contributions, your courage and your willingness to share.

## Acknowledgements

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**Thank you, scientific investigators.** Where would we be without your research and dedication to finding a cure for LAM?

**Thank you, everyone at NIH.** You've helped us become more knowledgeable about the disease while, during the original protocol, you pampered us with little vacations – and frequent-flyer miles! And if that weren't enough, you've given LAM patients a chance to build lasting friendships with each other and with the NIH staff.

Okay. Those were my general comments. Now I need to recognize the people who added real sweat and tears to this writing project. Although I can't name everyone who worked so hard on the handbook, I wish to thank certain people specifically. If you have helped and don't see your name, I sincerely apologize. But I do thank you nonetheless.

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# Chapter One:

## LAM 101 - The Basics



- 1.1 Lymphangiomyomatosis
- 1.2 The Epidemiology of LAM
- 1.3 Treatments for LAM



## 1.1 Lymphangiomyomatosis

Lymphangiomyomatosis (LAM) is a progressive lung disease that usually strikes women\* during the prime of their lives. Lymphangiomyomatosis is pronounced lim-fan-gee-o-ly-o-my-o-ma-to-sis. “Lymph” refers to the lymph vessels and “angio” refers to the blood vessels. “Leiomy” means smooth muscle, and “oma” is a tumor. The last part, “tosis,” refers to a disease condition.

The disease is characterized by an abnormal growth of smooth muscle cells. These cells invade the tissue of the lungs, including the airways, and blood and lymph vessels. Although these cells are not considered cancerous, they grow uncontrollably within the lungs. Over time, the cells form bundles, in the walls of the airways, blood and lymph vessels, causing them to become obstructed. The cells produce materials which break down tissue, causing the formation of cysts. The delicate architecture of the lungs is destroyed and airflow is blocked, preventing the delivery of oxygen to the rest of the body.

### How Is LAM Diagnosed?

LAM manifests itself in a wide variety of ways, so it is sometimes difficult to diagnose. Although von Stossel first described it in medical literature in 1937, the disease is quite rare. Many doctors are unfamiliar with LAM, so many patients go undiagnosed or are misdiagnosed before eventually learning that they have LAM. The difficulty of the diagnosis is compounded by the fact that a lot of the symptoms of LAM are the same as those for other diseases like asthma, emphysema, and bronchitis. Chest x-rays are not usually sufficient to detect LAM, but a high-resolution chest CT scan (HRCT), which shows the characteristic cystic structure of LAM, can usually provide an accurate diagnosis, if there are other manifestations of the disease (e.g., benign kidney tumor, lung collapse, or fluid in the lungs).

Additionally, a VEGF-D blood test may be used in combination with an HRCT to diagnose LAM. In some cases, an elevated VEGF-D level can indicate that LAM is present. However, there is also a high likelihood of a false-negative result. In some circumstances, a lung biopsy may need to be performed to provide an accurate diagnosis.

In addition to being difficult to diagnose, the cause of LAM is still unknown, and no cure is yet available. However, treatment of the disease with sirolimus (Rapamycin) has been approved by the FDA. (See section 1.4 – Treatments for LAM)

### What Happens in Lungs With LAM?

When you breathe, oxygen passes into the blood stream through tiny airspaces (alveoli) in the lungs into tiny blood vessels called capillaries. Normally, alveoli have walls that are only two cells thick. But as the smooth muscle cells of LAM multiply, these cell walls thicken, and oxygen can no longer pass through them efficiently. As it becomes harder for the oxygen to pass into your blood, your blood-oxygen levels will drop. That drop, in turn, forces your heart to work harder to pump oxygen to the various parts of your body. Your body tries to compensate by

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\* There are a few documented cases of men diagnosed with LAM, but LAM primarily affects women.

producing more red blood cells to deliver oxygen throughout the body. The growth of the cells also makes the structure of the lung more rigid. This loss of tissue flexibility causes the person with LAM to have difficulty moving air out of the lungs. This airflow obstruction makes you short of breath, a condition called dyspnea.

And if all of this weren't enough damage, this thickening also results in a breakdown of the lung tissue and the formation of small air-filled cavities, called cysts. As the disease progresses, there can be more extensive growth of smooth muscle cells throughout the lung and an increasing number of cysts are formed.

### **What Are the Symptoms?**

Many women with LAM suffer primarily from respiratory problems such as dyspnea (shortness of breath), chest pain, chronic cough and pneumothoraces (lung collapses). Some patients have benign kidney tumors called angiomyolipomas (AMLs) or an accumulation of milky fluid in the chest or abdomen called chyle. The abnormal cells that grow in the lung can block blood vessels in the lung, causing them to become swollen with blood and even to rupture, causing hemoptysis, the coughing up of blood or blood-stained sputum.

Not every woman who has LAM has all of these problems. About two-thirds will have at least one lung collapse (pneumothorax), and about one-third will have leakage of fluid into the chest cavity (chylothorax). Angiomyolipomas (benign kidney tumors), which are usually asymptomatic, are found in up to 50% of women with LAM.

Each of these problems is addressed in a separate chapter. We suggest that you first read the chapters that apply to you. As your expertise grows, you may want to read about other possible problems and treatments.

### **What Is the Course of LAM?**

We know that LAM is a progressive disease, but its progression is usually very slow with only tiny changes in breathing functions. Although it does not happen very often, women with LAM can progress at a rapid rate. Doctors are working to find indicators that will help to determine who will have a faster progression and who will progress more slowly. Whether disease progress is slow or rapid, lung function tends to decrease with time, and many LAM patients will, at some point, require oxygen therapy. Lung transplantation is often considered as a last resort. While many women with LAM add several years onto their lives through lung transplantation, it is not a cure.

## **What Can I Expect?**

LAM affects each woman quite differently. Some women have no symptoms while others have some of the problems mentioned above. Previously, the initial prognosis for a LAM patient was poor, but it was because LAM was generally only diagnosed in the advanced stages. Newer technologies such as the high-resolution CT scan have allowed for a much earlier diagnosis. In many cases, a woman's diagnosis has been accidental – for example, when a CT scan was done for a totally unrelated reason. Now investigators believe that many women who have LAM may never be diagnosed because their symptoms are too mild to bring them to medical attention.

Additionally, with the discovery of Rapamycin as an effective treatment for LAM, the prognosis for LAM patients has become much better. Rapamycin has been seen to slow disease progression for some LAM patients.

LAM patients have been followed at the National Heart, Lung, and Blood Institute (NHLBI) at the National Institutes of Health (NIH) since 1995, and researchers have learned more about how LAM affects each woman differently. Many women with LAM live a lot longer than previously predicted. Remember: the rate of progression and the incidence of resulting problems can—and does—vary considerably among patients.

While many questions about LAM have yet to be answered, the extraordinary progress in understanding the disease has provided real hope that a treatment and possibly a cure for LAM can be achieved.

## **How Is LAM Treated?**

Because LAM primarily strikes women, particularly those of childbearing age, physicians have thought that the hormone, estrogen, might be involved in the abnormal muscle cell growth that characterizes the disease, just as it is in the growth of smooth muscle in the uterus during a woman's childbearing years.

Although there is no direct evidence of a relationship between estrogen and LAM, in the past much of the treatment of LAM has focused on reducing the production and effect of estrogen. That's why women with LAM are sometimes advised against taking birth control pills. Even though there is no proof that having children will make your disease worse, there are case reports of LAM symptoms progressing during childbearing, and some doctors advise against pregnancy as it could accelerate the disease. More research still needs to be done in this area.

Two treatments that were commonly used in the last twenty years were the administration of medroxyprogesterone, a synthetic version of the hormone progesterone, and/or oophorectomy, the removal of the ovaries, to stop the body's main estrogen producer. Neither of these treatments has been scientifically proven. Because of a lack of proven benefit, the risks of surgery, and the possibility of future osteoporosis and heart disease, surgery to remove your ovaries is no longer recommended.

Rapamycin (also known as sirolimus) has been shown to slow the progression of LAM in some patients. On May 27, 2015, the FDA approved the use of Rapamycin for use in patients with LAM. A similar drug, called Afinitor (or everolimus) is also often used to treat patients with LAM and Tuberous Sclerosis Complex.

While these treatments can improve the lives of LAM patients and slow the progression of the disease, they are not a cure. The LAM Foundation continues to work diligently toward that goal.

### **What Research is Being Done?**

We know much more about LAM than we did just ten years ago! The LAM Foundation actively funds scientists to search for the cause of this disease and to ultimately find a cure. Dr. Elizabeth Henske, a LAM/TS researcher at Brigham and Women's Hospital in Boston, MA, discovered in 2000 that a mutation of the TSC2 gene (one of two genes responsible for tuberous sclerosis) is present in LAM cells, so LAM appears to be caused by a mutation of that gene.

Tuberous sclerosis (TS) is a hereditary neurological disease that has been linked with LAM. Recent screening of female TS patients has revealed that approximately 40% of them develop pulmonary LAM. Women who have LAM, but who do not have TS, are said to have "sporadic" LAM. At this time, researchers don't believe that sporadic LAM is hereditary. Mother-daughter transmission of sporadic LAM has never been reported. The similarities in the lung and kidney problems of sporadic LAM and TS are of major interest to researchers.

In addition to Henske's work, there has recently been rapid progress in understanding the pathways and the regulation of cell growth. This progress provides evidence that the mutations in TSC1 (which forms the protein, hamartin) and in TSC2 (which forms the protein, tuberin) result in the uncontrolled growth of LAM cells.

One of the big questions about LAM is whether the disease originates in one location in the body and then spreads to other locations (the metastatic theory) or whether it arises independently in several locations. In some patients who have undergone lung transplants, LAM cells from the recipient, not the donor, have been found in the donor lung. This finding supports the metastatic theory of LAM. If LAM cells are able to metastasize, then there could possibly be some treatments early in the disease – treatments that would be based on interfering with the migration of LAM cells.

The LAM Foundation is also working hard to fund research for projects that have a direct benefit to the patient. The LAM Biomarker Innovation Summit, a highly interactive two-day meeting produced recommendations for short and long term LAM biomarker development strategies. Over 45 LAM scientists and biomarker thought leaders joined together and presented current progress, discussed future priorities and provided recommendations.

The LAM community knows that identification of biomarkers will help improve therapies and also help women living with LAM to make more informed decisions throughout their lives. As a result of this summit, the following grants were issued and are currently being investigated.

### **Tuberous Sclerosis**

If you were also diagnosed with tuberous sclerosis complex (TSC) when you were diagnosed with LAM, don't be discouraged. You may have a milder form of TSC if you haven't had problems with it earlier. Many people refer to tuberous sclerosis as TSC, tuberous sclerosis complex. TSC is used frequently since TS also refers to Tourette's Syndrome.

Not all women with TSC have LAM, but more and more women with TSC are finding that they do have both diseases. This connection doesn't surprise doctors because, in both diseases, two genetic defects are seen in either the TSC1 gene (found on chromosome 9) or the TSC2 gene (found on chromosome 16). An abnormal copy of the gene exists in every cell of the body in tuberous sclerosis patients, causing the various manifestations of TSC that can encompass the entire body. Years ago, people believed that if someone had TSC that person would have three problems: mental retardation, skin growths on the face, and seizures. Now, doctors know that only 30-40% of people with TSC have all three of those manifestations.

Just as LAM affects women differently, TSC does as well. Although some people have severe forms of TSC, many live very normal lives. Severe cases of TSC may involve brain disorders including seizures, mental retardation, or developmental delays caused by the appearance of benign tumors in the brain. Quite often, these brain disorders show up in childhood, often leading to the TSC diagnosis. Fortunately, some of these brain disorders often end in childhood too.

In other people, TSC appears as tumors in internal organs, especially the kidneys (these can be angiomyolipomas, just as in LAM). TSC also affects the lungs with the same types of problems as LAM—shortness of breath and/or lung collapses. On a CT scan, TSC cells show a pattern almost identical to that of LAM cells. Some lucky people have even milder forms of TSC that show up as multiple lesions in the retinas of their eyes or fibromas (benign tumors made up mostly of fibrous tissue) on their faces that resemble acne. Even fingers and toes aren't exempt from the disease; they may have tumors on them that occasionally need to be removed.

If you or a loved one has TSC, you can find more detailed information about the disease by contacting the TS Alliance at 800-225-6872 or checking out the website at <http://www.TSAlliance.org>.

## **1.2 The Epidemiology of LAM**

LAM is a disease that occurs almost exclusively in women. A few cases of LAM have been reported in men, mostly those who also have another genetically-related disorder called tuberous sclerosis (TS).

All races are affected, and women with LAM have been identified in at least thirty-eight countries worldwide. The average age of women when diagnosed with LAM is about thirty-five, though, at the time of diagnosis, the reported age range is quite large—from twelve to seventy-

five years. Also, most women with LAM have had symptoms for three to five years before ultimately being diagnosed. Although LAM is considered to be a rare disease, physicians believe that it is not as rare as previously thought. Current estimates of the incidence of LAM are based on data from registries in the United States, France, and the United Kingdom, three countries where there have been substantial organized efforts in contacting pulmonary physicians to determine the numbers of patients with LAM. Because LAM is relatively uncommon, it's often misdiagnosed as asthma, bronchitis, emphysema or some other interstitial lung disease. Therefore, the current number of women diagnosed with LAM is very likely an underestimate of the true prevalence. Rough estimates suggest that there are 30,000 to 50,000 patients with sporadic LAM worldwide.

Some cases of LAM occur in patients who also have tuberous sclerosis or tuberous sclerosis complex (TSC- related LAM or TSC-LAM). It now appears that as many as 30%-40% of women with TSC may also be affected by LAM. The prevalence of TSC-related LAM is better established from population data. TSC is known to occur in approximately one of every 6,000 births, making the estimated number of TSC patients on earth about one to two million. Because TSC occurs equally in women and men, there may be 8,000-10,000 cases of TSC-related LAM in North America and up to 250,000 patients worldwide.

There are many similarities between sporadic LAM and TSC-LAM (e.g., the lung involvement and the benign kidney tumors), but there are also substantial differences. Women with TSC-LAM tend to have the same lung problems as those with sporadic LAM and can be just as severe. There have been reports of transmission of TSC-related LAM from mother to daughter (because TSC is an inherited disease), but there are no reports of the transmission of sporadic LAM.

### **1.3 Treatments for LAM**

Because of the amazing research done by our LAM scientists, today there is much more hope for the treatment of LAM than there has ever been! In the past, healthcare providers were only able to treat the many different symptoms of LAM. In 2015, the FDA approved the use of Rapamune for the treatment of LAM as an overall disease.

#### **Rapamune**

Rapamune is the first drug to be approved by the FDA for the treatment of LAM. Rapamune is the brand name of the drug but sometimes you'll hear it referred to as Rapamycin. This is the same drug. The generic name for the drug is sirolimus. It can be confusing but know that all of these names are referring to the same drug.

Rapamune has been shown to slow or even stop the progression of LAM for patients while they are taking the drug. Basically, it suppresses the growth of LAM cells and likely shrinks them without killing them. It has proven to be helpful in the lungs, in the kidneys and in the management of chyle leaks.

Unfortunately, the benefits of taking Rapamune do not come without some side effects. Some of the most common side effects are mouth and lip sores, diarrhea, abdominal pain, nausea, sore throat, acne, chest pain, leg swelling, upper respiratory tract infection, headache, dizziness, muscle pain and elevated cholesterol. Every person on the drug reacts a little

differently and therefore, their side effects are different. It's important to weigh the benefits you might receive from taking the drug with your side effects. There are many things you can do to help manage the side effects, such as taking a probiotic for intestinal symptoms or using a special mouthwash to help control mouth sores.

Because each patient metabolizes Rapamune differently, the dosage for each patient can be different. Instead of determining your dosage with a set amount, your pulmonologist will likely need to do frequent blood tests to look at the trough level of Rapamune in your body. It's important to communicate with your doctor regularly regarding your Rapamune dosage.

### **Afinitor**

Afinitor is another drug that is sometimes used for the treatment of LAM but has not been FDA approved for that purpose. It is, however, approved for the treatment of tuberous sclerosis and has been shown to be effective in the treatment of angiomyolipomas. The generic name for Afinitor is everolimus. This drug is in the same class as Rapamune and has similar functions and side effects.

Because the two drugs are so similar, if you are not able to take Rapamune, sometimes your doctor will give you Afinitor to try and vice-versa. Both have been effective for different patients and you should talk to your doctor about which drug would be most effective for you.



# Chapter Two:

## Diagnostic and Routine Tests



- 2.1 Diagnosing LAM
- 2.2 Pulmonary Function Tests
- 2.3 Imaging Tests
- 2.4 Cardiac Tests
- 2.5 Lab Work



## 2.1 Diagnosing LAM

LAM is not always easy to diagnose because it's rare, and also because it mimics other diseases like asthma, emphysema and bronchitis. If you were diagnosed recently without being misdiagnosed first, be thankful for that and be thankful to The LAM Foundation for its efforts to educate doctors and clinicians about the disease. If you were diagnosed years ago, your diagnosis probably took far longer to confirm. Nowadays, most cases of LAM can be diagnosed with a high-resolution CT scan (HRCT), especially if you have other symptoms, such as a lung collapse, shortness of breath or chest pain. Sometimes an angiomyolipoma, a benign kidney tumor, or a collection of fluid in the chest cavity (chylothorax) can help to establish the diagnosis.

A diagnostic blood test called VEGF-D can also be used in conjunction with a HRCT to distinguish LAM from other cystic lung diseases. In some cases, an elevated VEGF-D level can replace a biopsy to confirm diagnosis. This test may also be used on women with tuberous sclerosis to screen for LAM.

When other clinical clues are insufficient, a lung biopsy is often necessary. The most commonly performed procedure is a video-assisted thoracoscopic biopsy. But even now scientists are working to find other ways—like blood and urine tests—to diagnose LAM. If the progress continues, you may see these tests in the future. In the meantime, the following tests are the most current ways to diagnose LAM.

### CT Scan

A CT scan (also referred to as a CAT scan or computerized tomography scan) is an x-ray process in which a machine takes pictures of “slices,” “planes,” or “cross sections” of the body at specified intervals and from many angles. These images, which can be viewed on a monitor or reproduced as photographs, give the impression of looking into the body from the top instead of from the front, back, or side. CT scans look more three-dimensional than standard x-rays. The CT scans used to diagnose LAM are high-resolution scans. Basically, that means they take thinner sections and show more detail than standard CT scans. To diagnose or to establish the extent of LAM, you may receive a chest, abdominal, and/or pelvic scan. It is a good idea for LAM patients to obtain a head CT or MRI at least once in their lifetime to screen for tuberous sclerosis.

Before you have a CT scan, you must sign a consent form. The x-ray technician will want you to confirm that you're not pregnant and to indicate if you're allergic to the contrast medium the scan requires. If you do have allergies to that substance, the technician will proceed with the scan but without the use of contrast dye.

Even before you give your consent, you'll need to prepare a bit for the test. Usually, you'll fast for 4 to 6 hours if you'll be receiving contrast dye or if you feel you'll need to be sedated. Just before the procedure, you'll change into a hospital gown or you can just go to the test in clothing that has no metal pieces.

In preparation for the abdominal or pelvic scan, you may be asked to drink a barium solution that, when in your system, will provide greater visibility of the organs being scanned. The barium solution doesn't usually taste terrific (there are several flavors), but it's not too bad. You'll be asked to lie flat (usually on your back but sometimes on your stomach or on one side) on a narrow table that slides into the scanner. If you'll be having a scan with intravenous

contrast dye, an IV will be inserted into your arm before you're scanned or after the first set of scans are performed.

The scanner emits x-rays in a complete circle around you. Each slice of x-rays lasts a few seconds, then the table is advanced into the scanner a slight amount, and a new slice of x-rays is taken. During the test, you must remain still. A recorded voice will tell you when to hold your breath and when to breathe out. But don't worry: a sound system is built into the machine to allow for communication with the technician, a real person, during the test. If you have trouble holding your breath for the amount of time needed, let the technician know and the hold time will be adjusted.

At some point in the procedure, a contrast medium (a type of dye) may be injected into your vein to help define blood vessels and certain tissues, making them easier for the doctor to view. When the dye is injected, you may feel a "rush" or a warm sensation, which lasts only a few minutes. The entire procedure generally lasts between fifteen and thirty minutes.

If your doctor suspects LAM, he or she will examine your chest CT scan for evidence of the unique cystic pattern characteristic of LAM. If you have not had a CT scan of the abdominal area, you might want to suggest to your doctor that you have one so he or she can look for angiomyolipomas, benign kidney tumors. These and other clues will help in the diagnosis of LAM. Sometimes these are visible on the abdominal cuts of the chest CT, and a dedicated abdominal scan is unnecessary.

### **Bronchoscopy**

If the results of the CT scan aren't conclusive, a bronchoscopy may be considered for patients with puzzling lung disease. In general, however, the amount of tissue that is obtainable by this procedure is insufficient to make a definitive diagnosis of LAM and is not the preferred method for diagnosis. This test is also used, especially after lung transplants, to check for signs of infection or rejection.

A bronchoscopy is an examination of the inside of your lungs. Your pulmonologist performs the examination by guiding a bronchoscope, a flexible tube with a fiberoptic imaging attachment on the end, down into your lungs to examine the airways. Usually the scope goes down via one of your nostrils, then down through your trachea (windpipe), and finally into your lungs. When your pulmonologist extracts a small amount of lung tissue for biopsy during the bronchoscopy, it's called a transbronchial biopsy. The extraction is sometimes done with tiny forceps or by using bronchial lavage, that is, introducing a tiny bit of saline solution into the lungs and suctioning it back out.

Prior to the bronchoscopy, which can be done as an outpatient procedure, you'll usually have some blood work, pulmonary function tests (PFTs), and a chest x-ray. Generally, you'll be asked to fast before a bronchoscopy, so you won't be allowed anything to eat or drink the night before the test.

When you get to the hospital or clinic, an IV will be started. Once that is inserted, you'll inhale a mist of Lidocaine, or some other numbing solution, to numb the mucous membranes in your nose and throat. Sometimes you'll also be given Lidocaine to rinse and gargle with. You want your throat to be very numb before the tube is inserted to eliminate the gag reflex that occurs when an object is placed into the back of your mouth.

Once your throat is numb, a nurse will swab even more Lidocaine in your throat and will put Lidocaine gel in your nose. You'll be given a medication like Demerol or Versed to relax you. Just before the procedure, you'll be hooked up to an EKG monitor, and a pulse oximeter will be placed on your finger. You may also have oxygen set up to flow through the nostril that's not used.

Generally, because of the drugs, you'll probably not be fully alert during the procedure, but if you are you may feel liquid trickling down the back of your throat and you may feel the need to cough. If you do cough, you'll receive more anesthetic.

When the procedure is complete, you'll be monitored until the drugs wear off and until you're again breathing comfortably on your own. Once your coughing reflex returns, a signal that your throat is no longer numb, you'll be allowed to eat and drink. Another chest x-ray may be done to insure that you didn't suffer a small pneumothorax (lung collapse) or some other complication during the procedure. There's also a slight chance that you'll need to stay overnight for observation.

### **Lung Biopsy**

Sometimes a thoracoscopy is used to perform a lung biopsy to diagnose LAM. This operation requires general anesthesia, and obtains a tissue sample through a scope rather than a large incision in the chest wall. This procedure is less painful and usually has a shorter hospital stay and recovery time. For more information on thoracotomy and thoracoscopy, see the section on the management of pneumothorax.

A thoracotomy is a major procedure in which a surgical incision is made in your chest wall between your ribs. While you're under a general anesthesia, the surgeon makes an incision in the chest, an incision that allows the doctor access to lung tissue for a biopsy.

This procedure is an invasive surgery, so depending on the size of the incision and the length of the procedure, you'll require anywhere from one night to a weeklong stay in the hospital. Full recovery can take anywhere from a few weeks to a few months. Because of the invasiveness of the procedure, it should only be performed if extremely necessary.

Once you receive your official diagnosis, you'll undergo many other tests—from blood work to pulmonary function tests—to evaluate your disease and to monitor its progression. Many of these tests and procedures are described in the following sections.

## **2.2 Pulmonary Function Tests**

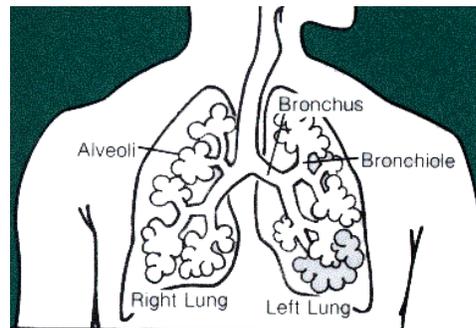
Pulmonary function tests (PFTs) evaluate how well your lungs are working. They can reveal the presence of lung disease or an abnormality of your lung function. The pattern of changes from normal values can diagnose certain types of disorders and evaluate their severity. PFTs provide an objective measurement of your lung function rather than a subjective evaluation—like your opinion—of the severity of your own symptoms. But in general, how you feel is related to how normal or abnormal your PFTs are.

Pulmonary function tests don't measure the disease itself or the number or size of cysts in your lungs. However, when your PFTs are compared to those of a reference or control population, the actual test results take on more meaning and give the doctor more information.

A reference population is a group of people who are the same age, sex, height, and ethnicity as the person taking the test. The test scores of these people are averaged. This average appears as the *predicted value* (or *reference value*) on a test, and that is the number your score is compared to. In addition to comparing your scores to those of the reference group, your doctor needs you to take multiple sets of PFTs over a period of time to determine if your disease is progressing. Since there is always a margin of error in these tests, multiple sets of tests taken together indicate whether there is a progression of your disease.

### **The Lungs and the Mechanics of Breathing**

To understand how LAM affects you, you need to have a basic understanding of the lungs and their structure. Your lungs are two sponge-like organs. Your right lung is made up of three lobes or sections, and it's a bit larger than your left lung, which has only two lobes. These two organs are connected by bronchial tubes. Think of a tree upside-down. Your trachea, also called the windpipe, (a tube that leads downward from the larynx or voice box) is equivalent to the trunk of the tree. The trachea branches off into two main sections or bronchi (the plural of bronchus): the left bronchus and the right bronchus, which extend into the lungs. Branching out from the bronchi are the bronchioles, a smaller series of tubes. As the airway structure divides more and more, these sections become smaller and smaller, just as the branches of a tree become smaller the higher up they are in the tree. The bronchioles end in tiny air sacs called alveoli (singular: alveolus). These tiny sacs have thin cell walls that are surrounded by tiny blood vessels. Because alveolar cell walls are so thin, they're able to pass oxygen into the blood. In exchange, carbon dioxide, a waste product of your body, goes into the sacs, up the bronchioles, to the bronchi, up the trachea, and finally is breathed out of the body.



Source: [www.cancer.gov/cancertopics/wyntk/lung/page3](http://www.cancer.gov/cancertopics/wyntk/lung/page3)

A brief explanation of the mechanics of breathing can give you a better idea of the problems caused by LAM. The diaphragm is a large muscle under the lungs. In normal lungs, the diaphragm has an upwardly curved shape when the lungs are at rest. As you inhale, the diaphragm contracts downward along with the muscles attached to the ribs. These movements cause the chest cavity to enlarge, pulling air into the lungs. The elasticity of the lungs causes the diaphragm and rib muscles to recoil, making the chest cavity smaller and pushing the air out of the lungs. You exhale, deflating the lungs. This is the normal pattern of inhaling and exhaling.

With lung diseases like LAM, this elasticity is often reduced due to the growth of LAM cells. The reduction in elasticity often causes air trapping, a problem that increases the lung volume. When the lung volume is increased because of air trapping, the diaphragm is forced to flatten and cannot recoil and push air out as readily as it did before.

Another feature of normal breathing occurs in the small airways. In healthy lungs, these small airways increase and decrease in size with inspiration and expiration. Elastic tendons tug on the alveolar\* walls of the airways and pull them open as you inhale. When air passes through, the tendons will release and the airways close. In lungs with LAM, the proliferation of smooth muscle tissue decreases the elasticity of the tendons. The tendons can't open the small airways as well as they did previously, and the loss of elasticity also causes the airways to close prematurely and obstruct the airflow. When this happens, gas becomes trapped, the lungs become larger due to the trapped air, and the diaphragm flattens and doesn't work as well. Your scores on the PFTs measure all these changes in the ability of your lungs to work normally.

### **Performing Pulmonary Function Tests**

Pulmonary function tests (PFTs) are usually performed in a pulmonary function laboratory. During the test, you'll sit in a chair and wear a clip on your nose while breathing into a tube connected to a machine that will record the test results. The tests are painless, but you may find them fatiguing.

Often, after you have performed the different tests, you'll be given a bronchodilator, a drug that allows your airways to open (relax) more, like Albuterol to inhale. You'll then be asked to repeat several of the tests to see if your breathing improves after using this drug. Not all women with LAM see improvement, but some do.

### **Accuracy**

The first time you perform a set of PFTs you might wonder if your scores are accurate. In general, they're usually accurate, plus or minus a few percentage points—but not the first few times you take the tests. Learning how to breathe correctly during PFTs takes practice, so your first few sets may not be as accurate as subsequent ones. Your test results, however, may vary on any given day, and that's why repeated tests are so useful.

Accuracy depends on several factors including your effort, the skill of your technician, and the machine, called a spirometer, that you're using for the test. Hence, some of the tests are called spirometry.

To get the most accurate scores, wear loose clothes that don't restrict your breathing, and don't eat too much before a test. If your stomach is too full, it may press on your diaphragm, thereby limiting your ability to breathe deeply. If you use inhaled medications such as bronchodilators, your doctor or technician may ask that you refrain from taking those before the test so you can get a more accurate measurement.

When you perform these tests, you'll be asked to repeat each part of the test at least three times. Your expirations should meet "acceptable" standards and be reproducible.\* Repeating a test several times allows your technician to get a more accurate evaluation of your overall performance, insuring that a very high or a very low score is not a fluke. Performing every test at your maximum effort ensures the best results. But always keep one point in mind: Your PFT results may not be totally accurate on any given day. As mentioned, PFTs always have a percentage of error, and sometimes you may not perform well due to a cold, allergies, fatigue, or even the time of day. But, because you'll be doing these tests on a regular basis, a trend will

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\* Alveoli are tiny air sacs at the ends of bronchial tubes in the lungs where gas exchange occurs.

\* The American Thoracic Society has sets of rules for your technician to follow. You'll need to repeat the tests according to these guidelines.

appear after a few series of tests so that you'll be able to learn if you're stable or if the disease is progressing. One day you may have a frighteningly low score on a section of the test, but six months later when you take the test again, your score may be more consistent with your previous results. You have to condition yourself not to get upset about one bad score or one bad day.

You'll also find that the respiratory technician who administers your tests is vital to performing a test accurately and successfully. A good technician will explain what you'll be doing before you breathe into the machine and will also coach you through the test. Very few people can remember what to do once they put on nose clips and place their mouths around the mouthpieces. If you're able, it can be helpful to request the same technician each time because, as you learn that person's techniques, you'll receive better results. Plus, you'll probably feel more relaxed another factor which contributes to the success of the testing.

The last factor in good test results is the machine you're using. In a good lab, the machines are checked regularly and are calibrated accurately. A well-maintained machine will give you true results. You might, however, notice a slight difference in your scores done on machines in your local lab compared to the machines at The National Institutes of Health if you're in the protocol there. Slightly different tests scores on different machines are quite normal, but your test results should be relatively close. Different institutions may use different reference equations to figure percent predicted, so when comparing results from one institution to another, look at the actual or measured values not the percentages.

The American Thoracic Society (ATS) dictates how these PFTs should be done, the proper standards for accuracy, the specifications of the equipment and the sanitary practices to ensure safe testing. The standards also state that each patient should exert her maximum effort, so you may see your technician recording what he or she perceives your effort to be. These guidelines also dictate that three attempts must be acceptable and, of those three, two of them need to be reproducible (within a certain % of each other) to be considered accurate. Obviously, sometimes this is not possible, so the best results are used.

## **The Scores**

Your test results indicate to your doctor and to you how serious your lung disease is and whether your disease is progressing. When you look at your tests, you may be overwhelmed at all the different values and scores, but some values are more important than others, and those are the ones you need to focus on.

First of all, remember that your scores are compared to predicted values (average numbers of a reference population) based on age, sex, height, and ethnicity.\* In general, a normal score would be considered as 80% to 120% of the predicted value.

PFTs are divided into three main sets of tests: those that measure function (the mechanics of breathing) and airflow (spirometry), those that check lung volumes, and those that look at your diffusing capacity, that is, the exchange of gases to and from your respiratory system and your circulatory system. PFTs are also used to assess response to a bronchodilator. Some doctors order a full set of PFTs for each of your visits, but some may just ask for the full set once a year or so and just use spirometry for interim visits.

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\* Although you may find it surprising, some people actually volunteer to perform these tests just to act as controls for scientific comparison.

## Spirometry

As mentioned above, spirometry is a test that provides measurements of airflow (or mechanics of breathing). It's performed on a machine called a spirometer. This test measures how much air you can inhale and exhale. It also measures how fast you can blow out air. There are two values in spirometry—the forced expiratory volume in one second (FEV1), and the forced vital capacity (FVC). The FEV1 is one of the most important values for LAM patients. But the FVC and the ratio of the two values, FEV1/FVC, also yield important information.

The maneuver for spirometry is as follows:

Spirometry: Normal breathing. Take a big breath in and blow it out as fast and as long as you can. Keep blowing until you're empty and can't blow out any more air. Keep blowing. Keep going. Take a big breath in.

The FEV1 is the maximum amount of air you can forcibly blow out in one second after a full inhalation. This test measures obstruction of airflow in the airways. As your lung tissue loses its elasticity, your airways shut prematurely. When this happens, you sometimes exhale less air or it takes a lot longer for you to exhale than it does for a normal person. In LAM, the FEV1 is usually reduced. The rate of decline in FEV1 is an index of the disease's progression. Normal people lose about 10-25 ml/year of FEV1. LAM patients lose an average of about 100 ml/year, and smokers lose about 70 ml/year.

In the FEV1, the amount of obstruction determines the stage of severity of LAM. These numbers are compared to the reference population. The following percentages are a "rule of thumb" for defining severity. Other doctors might use different percentages and other factors to rate your severity.

Mild:	< 80% predicted
Moderate:	< 70% predicted
Moderately Severe:	< 60% predicted
Severe:	< 50% predicted

Don't just look at the percentage of predicted. The actual liter amount you blow out is usually a better indicator.

The FVC is the total amount of air you can exhale forcefully in the spirometry maneuver. This is another indicator of obstruction. In LAM, the FVC is usually normal or reduced.

The FEV1/FVC result is not a value in itself, but the ratio of the FEV1/FVC. It is also a very significant number. Your actual numbers from the FEV1 and the FVC are used. This ratio is expressed as a percentage (70-85% is considered normal for an adult) and a reduction in this number usually indicates obstruction of the larger airways, a condition where you have trouble blowing air out of your lungs. Sometimes, below normal FEV1 and FVC measurements yield a normal to high ratio. This result is usually an indication of restrictive disease. In women with LAM, restriction, or being unable to get enough air in, is less common than obstruction. However, sometimes it feels as if you can't get enough air in, but this feeling is due to the inability to exhale efficiently.

There are two other values for spirometry. The FEF25-75 refers to how fast you can blow out in the middle 25-75% of the test. The FEF25-75 measures the mid to smaller airways (peripheral airways), and can be an early indicator of airway obstruction.

The other value, the PEF, Peak Expiratory Flow Rate—also called the Peak Expiratory Flow (PEF), the Peak Flow (PF), or the Forced Expiratory Flow Maximum (FEFMax)—indicates the fastest speed you blow out when you’re performing the FVC maneuver. Because it’s more influenced by your effort than other tests, it isn’t as strong an indicator of problems as the other tests.

Spirometry is also used to assess response to a bronchodilator. After you perform your PFTs, you may be given a bronchodilator to inhale. If improvement is seen after using the bronchodilator, there is some responsiveness of your airways (increase in lung function). Then you’ll probably be started on inhalers. If no improvement is seen after using a bronchodilator, it doesn’t mean that the bronchodilator won’t help; it may help with symptoms (e.g., decreasing your shortness of breath and/or your cough) or sometimes it may boost your peace of mind. In such cases, inhalers may still be helpful.

Your spirometry scores might look something like this.

	Actual	Predicted	% Predicted	Actual	% Post	% Change
FVC (L)	4.08	3.77	108	4.21	111	3
FEV1 (L)	2.03	3.00	67	2.54	85	25

With these results your FEV1/FVC would be 50%.

### How to Interpret the Results

Your airflow is measured in liters (L). Then the liter flow is calculated in percentages by comparison to the control numbers.

On the left side of the chart: your results without drugs

Actual The amount of air in liters (L) you exhale (or measured):

Predicted: The average response of the control group

% Predicted How you compared to the control group (% Reference):

On the right side of the chart: the results after you inhale a bronchodilator

Actual: The amount of air in liters (L) you exhale

% Post: How you compared to the control group after taking a bronchodilator

% Change: How much your flow changed or how much a bronchodilator helped you

As mentioned earlier, some calculations include ethnicity, sex, height and even weight in the reference population. Weight can have an effect on your ability to take a deep breath, but it has little effect on spirometry. But excess weight on anyone can make that person's heart *and lungs* work harder.

The person who performs your spirometry is very essential in helping you to achieve the most accurate test scores you can. He or she needs to be reliable and needs to motivate you and talk you through the tests so that you do your very best. PFTs can be very tiring and you can easily forget when you're supposed to inhale, exhale, or hold your breath. A good respiratory technician will constantly give you feedback and suggest tips for you to perform the tests as successfully as possible. If you feel you aren't being helped sufficiently, ask for help. Tell the person to remind you about what you need to do.

## **Lung Volumes**

How much air you move in and out of your lungs and how much air gets trapped in the lungs are essential questions that your doctors need answers to, so here are some other things they look at when evaluating your breathing. First, there are four lung volumes:

- The inspiratory reserve volume (IRV) is the amount of air that you can take in from a normal ("tidal" in doctor terms) resting inspiration to the fullest inspiration. To get this value, first you take in a normal or regular breath. At the end of that inhalation, you then take in as much extra air as possible.
- The expiratory reserve volume (ERV) is the extra amount of air you can forcibly exhale after you've exhaled normally.
- The tidal volume (TV) is the amount of air you inhale and exhale during your resting or normal breathing pattern.
- The residual or reserve volume (RV) is the amount of air left in your lungs after a full expiration, that is, after you've blown out as much air as you possibly can. (In LAM, the RV is usually increased because of trapped gas. The result of the air trapping is that you have less space to breathe air into.)

Combinations of these volumes make up lung capacities:\*

- The functional residual capacity (FRC) measures the volume of gas in the lungs at rest. Another way of saying this is that the FRC is the amount of air that is not expelled but remains in your lungs after a normal exhalation. This capacity is made up of two lung volumes, expiratory reserve volume (ERV) and the residual volume (RV). Once the FRC is known, the TLC (total lung capacity) and the RV can be determined. The FRC is measured by one of the following stated lung volume techniques.
- The inspiratory capacity (IC) is the total amount of air that you can draw into your lungs after a normal exhalation.
- The total lung capacity (TLC) indicates the maximum amount of gas the lungs can hold after you've inhaled as much air as you possibly can. In LAM, the TLC is usually normal or slightly increased because your lungs become hyper-inflated.
- The vital capacity (VC) is the amount of air that you can forcibly exhale after a full inspiration.

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\* Ask your doctor or respiratory technician for more detailed explanations.

Lung volume measurements can be obtained by one of three different methods: nitrogen washout, helium dilution, or body plethysmography (body box).

- Nitrogen washout: You breathe 100% oxygen for approximately seven minutes. Each time you exhale, you breathe out some of the nitrogen, which is in your lungs from your normal breathing. As you breathe in the pure oxygen, the nitrogen is measured with each exhalation until it is down to a certain level or until you can't perform the test any more. Once you "washout," that is, get the nitrogen out of your system, the FRC is calculated.
- Helium dilution: You breathe a known mixture of gas (usually with 10% helium) for approximately 2-5 minutes. As you're breathing on the system, the helium mixes with the gases in your lungs. Once "diluted," the system can calculate your FRC.
- Body plethysmography (body box): The technician can also calculate the volume of air in your chest by having you sit in a large glass chamber or box, known as a plethysmograph. When the door is closed and when the pressure in the box stabilizes, you'll be asked to breathe in different ways. While you're breathing, pressure readings are taken, readings that allow the computer to calculate the volume of air in your chest. This is the most accurate method for obtaining lung volumes and capacities.

	Actual	Predicted	% Predicted
TLC (L)	5.38	5.27	102
FRC (L)	2.47	2.70	91
RV (L)	1.28	1.50	85

Normal is considered to be 80-120% of predicted.

## Diffusion

The diffusion capacity for carbon dioxide (DLCO) checks your gas exchange: how well oxygen gets from the lungs (aveoli) into the blood stream and how well carbon dioxide gets from the capillaries into the lungs. A small measured amount (not enough to harm you) of carbon monoxide (CO) is inhaled into the lungs. You hold your breath for a short period of time. When you blow out, the machine measures how much CO you blow back out. If you blow too much CO back out, your capacity to pass oxygen into the bloodstream is reduced. In LAM, most patients have a reduced DLCO. This, like the FEV1, is a major indicator of the severity of the disease. But, as in all these tests, isolated readings can vary. It is important to watch for trends.

The maneuver for the DLCO:

DLCO: Normal breathing. Take a big breath in and blow it out slowly and completely. Take in another quick full breath. Hold it for about 7-10 seconds. Blow out again quickly and completely.

You may never fully understand all of your PFT values. If that's the case, don't be upset. Many women with LAM don't understand them. For those with LAM, the FEV1 and the DLCO seem to be the most critical values. Try to understand those, and let your doctors take care of the rest.

## 2.3 Imaging Tests

### Chest X-ray

A chest x-ray is often used to detect problems (e.g., pneumothorax and chylothorax) and abnormalities in the lungs, heart, and bones in the chest. If you go to the National Institutes of Health (NIH) for the LAM protocol, you'll get a chest x-ray each time you visit. LAM patients are usually x-rayed from two different directions. Once from the back, giving a view of the chest from the back to the front, and once laterally, that is from one side to the other.

The test is simple and painless. As with other types of x-rays, when you have a chest x-ray, you'll be asked whether you're pregnant. If you are pregnant, your doctor will have to decide if an x-ray at that particular time is safe for you. If you're not pregnant, you'll remove all of your clothes and jewelry from the chest and neck area and change into a plain surgical-type top or hospital gown.

A chest x-ray isn't usually sufficient to show the cystic structure of LAM, but it can indicate pneumothoraces and areas of pleural effusion and chylothorax.

### CT Scan

CT scans are often sufficient to diagnosis LAM, but they're also regularly taken to monitor the course of the disease. Please see the section on *Diagnosing LAM*.

### Magnetic Resonance Imaging

Magnetic Resonance Imaging (MRI) is a sophisticated imaging technique that uses magnetism and radio waves, rather than potentially harmful x-rays, to view internal structures of the body, especially soft tissues and the brain. The resulting images are very clear and quite detailed.

An MRI works like this: a powerful magnet pulls atoms toward it temporarily. Once the atoms are lined up with this magnetic field, radio waves are bounced at the nuclei of the atoms to disrupt them. The computer that is attached to the machine processes the energy given off by the nuclei as they return to their normal state, and then it interprets that energy as a picture or scan.

Because such strong magnetism is used, you'll need to remove any metal jewelry, zippers, or buttons. Your clothes and other belongings must be left in another room to protect them from the magnetic field, which can damage your credit cards and cell phone. But other than removing any metal you're wearing, no preparation is needed for an MRI.

During the procedure, you'll lie on a table that slides into a tubular scanner that is actually a large magnet. MRIs are known for their noise and somewhat tight space, but the newer scanners are quieter and less claustrophobic than the older models. And the scans take less time than they used to. Some technicians will offer you a choice of music to listen to during the test, but all technicians will offer you earplugs or other protection for your ears. If they don't offer, don't hesitate to ask for something to muffle the noise.

Except for the noise, the scans are quick and easy, and you won't need any recovery time unless you fall asleep during the procedure.

## **Ultrasound**

An ultrasound is another aid to assist physicians and technologists in viewing internal organs. A small hand-held instrument called a transducer bounces high-frequency sound waves, which the human ear cannot hear, off the organ being examined. The sound waves, which echo back, are picked up by the same device, which then sends the signals to a computer. The computer translates the impulses into an image on a monitor. Since the ultrasound is constantly emitting and picking up sound waves, the images are always moving; they aren't stationary like x-rays or MRIs.

Some women are familiar with prenatal ultrasounds that monitor fetuses during pregnancies. Other women may have had ultrasounds of their breasts to study suspicious lumps that show up during mammograms. The same ultrasound principles are used in women with LAM to check for angiomyolipomas (AMLs) in the kidneys and in other internal organs.

The technician performing the test will apply a lubricating gel to the area being scanned (kind technicians warm the gel first). The gel allows for good contact between the transducer and your skin, and it also allows the transducer to glide easily across your abdomen. You'll feel some pressure as the technician pushes the transducer into your abdomen to get as close as possible to an organ to get a good image. Every now and then, the technician will ask you to hold your breath so that a better scan can be taken.

Generally you don't need to prepare for an ultrasound, but occasionally you will be asked to fast so that your stomach, bladder or bowel isn't full and doesn't block the view of another organ. For some scans, like scans of the bladder, you may need to drink a large amount of water and not urinate until the test is over. But, generally, ultrasounds are relatively easy tests to undergo.

It is advised to wear loose-fitting clothes for an ultrasound, or change into a hospital gown because the gel can get on your clothing very easily, and although it washes out of your clothes, you'll feel damp and sticky until you change.

## **Bone-density Scans**

Some women with LAM suffer from osteoporosis, so they need to monitor this disease as well. These scans are the easiest scans of all. When you first get diagnosed, you should have this done to get a baseline scan, a scan against which all future scans can be compared. This scan will show the condition of your bones—whether you already have some form of osteopenia or osteoporosis, or whether your bones are still strong and dense.

If the scans show that your bones are fine, continue to monitor them with regular scans about a year apart or as often as your doctor recommends. If you're in the protocol at NIH, you'll receive a bone scan yearly.

Although there's no real preparation for these scans except to change into clothing without metal, you'll again be asked if you're pregnant and you'll probably sign a consent form. There is, however, one caution. If you're having a series of tests done, you must have your bone density performed before you're injected with any type of contrast dye, (e.g., during a CT scan). If you have the dye in your system, the bone scan will be difficult if not impossible to read.

Once you're ready for the scan, you'll be asked to sit or lie in several different positions. Several different parts of your body are scanned to get a better sense of your bone density. For a scan of your spine, you'll probably lie on your back with your legs resting on a cube, so that

your back will flatten out onto the table. You just lie still while the scanning machine moves around you automatically. If your hip is scanned, you'll lie flat, and your feet will be gently strapped to a board to hold them steady. For your wrist, you'll sit up, and rest your arm on the table; your technician will position your wrist. Generally, the positioning takes more time than the scan, but nothing is uncomfortable.

## 2.4 Cardiac Tests

Breathing difficulties can affect much more than your lungs. They can affect your other organs tremendously, especially your heart. Consequently, even though LAM is considered a lung disease, you should have your heart checked regularly.

### Blood Pressure

Almost every woman who's ever gone to a doctor has had her blood pressure read. It's probably the most common monitoring test there is, except perhaps taking your temperature. You might think that the following paragraphs devoted to blood pressure testing are excessive for such a simple test, but hypertension (the medical term for high blood pressure) is a very, very serious problem that must be detected and treated. Hypertension can cause heart problems, kidney problems and/or stroke and death. If you have a family history of hypertension, it's even more vital to have your blood pressure checked regularly.

Having your blood pressure monitored is very simple. A cuff with an inflatable bladder or sac inside is wrapped around your upper arm. This cuff is gradually inflated and deflated, either with an automatic cuff or manually. When the cuff is inflated, the air blocks the flow of blood that flows through a large artery. Deflating the cuff allows the blood to flow again. When the blood resumes its flow, a dial, which is attached to the cuff, registers the first heart beat (systolic pressure) and the last beat (diastolic pressure), which your nurse or doctor listens to with a stethoscope. Both the systolic and diastolic numbers indicate your blood pressure, for example, 120/80, with the upper number indicating the systolic and the lower number representing the diastolic.

To get a more accurate reading, don't drink coffee or other high-caffeine beverages beforehand. Make sure you have gone to the bathroom and emptied your bladder beforehand also. Sit with your feet flat on the floor, relax, and breathe normally. Clothing that allows access to your upper arm is easier on both you and the person taking your blood pressure.

According to the American Heart Association's literature,<sup>\*</sup> the explanation of blood pressure is as follows:

*When your heart beats, it pumps blood through large blood vessels called arteries. They conduct blood from your heart to other parts of your body. As your blood is pumped through your arteries, it pushes against the artery walls.*

*This force against the artery walls is called blood pressure.*

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<sup>\*</sup> All quoted sections are from a pamphlet "About High Blood Pressure", published in 1993 by the American Heart Association. You can get more information at their website: <http://www.americanheart.org>.

A healthy person's arteries are muscular and elastic. They stretch when the heart pumps blood through them; the amount they stretch depends on how much force the blood exerts. Your heart beats about 60 to 80 times a minute under normal conditions. With each beat, it sends a surge of blood into your arteries. That makes the blood pressure in your arteries go up. Conversely, when your heart relaxes between beats, your blood pressure will go down.

This means that you really have two levels of blood pressure. The higher level (systolic pressure) occurs when your heart is beating. The lower one (diastolic pressure) occurs when your heart is resting.

Both levels are measured which is why the reading is stated as *one number over another*. “The systolic pressure tells the maximum amount of pressure exerted on your arteries. The diastolic pressure tells the minimum pressure on your arteries. The harder it is for the blood to flow through your blood vessels, the higher both numbers will be—and the more strain on your heart.”

Recently, new guidelines for blood pressure in all adults 18 and older have been established.

Normal: <120/<80

Prehypertension: 120-139/80-89

Stage 1 hypertension: 140-159/90-99

Stage 2 hypertension: 160/100 or greater

If you have prehypertension or Stage 1 or 2 hypertension, your doctor will advise you about possible treatment or monitoring.

A condition called *White Coat Hypertension* —a reaction to the doctor's white coat—was touted as a reason for having a high reading in the doctor's office. This may be the cause of a high reading, but it's not dismissed as it once was. Monitoring over time will give a truer picture. If this reaction is a problem for you, ask to have your blood pressure taken again before you leave.

What can you do to help reduce blood pressure?

- Maintain a normal weight.
- Eat a diet rich in fruits, vegetables, and low-fat dairy products.
- Limit amounts of saturated fat and sodium (salt).
- Get at least 30 minutes of aerobic exercise per day.
- Limit consumption of alcohol to one drink per day.
- STOP SMOKING!

If you are prescribed medication to control your hypertension, be sure to take it according to the doctor's instructions. NEVER just stop because your BP is lowered UNLESS your doctor tells you it is OK.

## **Electrocardiogram**

An electrocardiogram (EKG or ECG) is used to measure the electrical activity of the heart muscle. To monitor the heart, electrodes are placed on your chest and extremities. These electrodes pick up the electrical impulses your heart emits and sends them to a computer. The results print out onto a moving piece of paper. The technician or your doctor will be able to tell whether the electrical activity in your heart is normal, too fast, or too slow. Other abnormalities can also show up.

During the test, you'll need to lie flat for about five minutes and perfectly still for about 10-20 seconds. Generally, no preparation is needed, but oily skin or lotion on your body may prevent the electrodes from making good contact with your skin. If that happens, the technician will wipe off small sections of your skin with an alcohol swab to ensure good contact.

## **Echocardiogram**

An echocardiogram is basically an ultrasound of your heart (see Ultrasound under the Imaging section) that allows a technician to determine the passage of blood through your heart. To get good results, the technician will apply a gel lubricant to your chest to allow the ultrasound sensor, called a transducer, to make good contact with your skin and to slide easily across it.

You may experience slight discomfort when the technician presses hard on your breastbone or ribs with the transducer, but the discomfort only lasts a minute. During the procedure, the technician looks at a screen or monitor, showing a two-dimensional image of your heart.

No preparation is needed for this test, but be warned: the ultrasound gel can be cold!

## **2.5 Lab Work**

Lab work is a necessity in monitoring any disease. Many of the tests your doctors request are routine ones, but your doctors will probably start to pay much more attention to every little change in your tests results—just to keep you as healthy as possible.

### **Blood Tests**

What tests will your doctor order? Cholesterol tests are the most common. The total cholesterol, the HDL (high-density lipoprotein) or “good” cholesterol, and the LDL (low-density lipoprotein) or “bad” cholesterol are all important as well as your triglyceride (neutral fats) levels. In addition to these, your doctor will probably monitor your thyroid function.

Most likely, a complete blood count (CBC) will be done. This shows the components in your blood—for example, white blood cells, red blood cells, and hemoglobin—as compared to a normal range. Abnormalities in these tests may indicate such things as infection or anemia.

A Chem Panel would also probably be one of the tests done. This panel shows the levels of common components such as sodium (salt), glucose (sugar), calcium, bilirubin (an indication of liver function), and potassium, among others. If you participate in the National Institutes of Health (NIH) protocol, there are between 25-35 different tests done. In addition, there are some tests that are specific to the LAM protocol.

Some of these tests, like the one for cholesterol, will require you to fast for 8-12 hours. Others won't require any preparation.

When a blood sample is needed, you'll usually have a venal blood draw, that is, taking blood from a vein, most commonly a vein on the inside of your elbow. The procedure is relatively painless, but, sometimes, the veins of people who have frequent blood draws become smaller, making future draws more difficult.

If you're having frequent blood tests, ask your nurse or phlebotomist (a person who draws blood for analysis) to use a smaller gauge needle. Sometimes the smaller gauge makes just enough of a difference to ease the procedure.

### **The 24-hour Urine Collection**

If you participate in the NIH protocol, you're already familiar with this test: the 24-hour urine collection. The title explains the test fully; you collect your urine for 24 hours.

The test is painless, but a bit awkward the first few times. You'll be given a plastic receptacle or "hat" to place on your toilet, right under the seat. It's shaped to collect your urine and also to allow your stools to bypass the hat entirely. Once you finish urinating, you'll pour your urine into a special container (perhaps even a simple gallon jug), and then refrigerate it until the 24 hours are over. Don't forget to wash your hands!

If you're doing this at NIH, your first morning urine will be checked to see if you're pregnant. Even if you know you're not expecting, even if you no longer have a uterus, the test will be performed. After that, each time you urinate you'll collect the full amount. The time of your first urination is noted, and you'll be woken up at that same time the next day—whether you have the urge to go or not.

So why are you doing this? There are several reasons. The first and most important reason is to see how well your kidneys are functioning. Good kidney function is essential because the kidneys purify your blood by processing the toxins that travel through your body. Not only do the kidneys purify your blood, but they also help to regulate your blood pressure. Additionally, the health of your kidneys gives your doctor an overview of your health.

Here are just a few of the things your doctor will be looking for in your urine:

- pH balance (i.e., how acidic it is),
- presence of blood or dangerous bacteria,
- concentration,
- glucose level.

As you can see by what a urine test can tell you, the kidneys are vital organs all the time. But they're even more critical after a lung transplant. Then the kidneys are needed to process all of the powerful immunosuppressants that are taken.

# Chapter Three:

## Taking Care of Yourself



- 3.1 Coping with Your Diagnosis
- 3.2 Pulmonary Rehabilitation
- 3.3 Borg Rating of Perceived Exertion Scale
- 3.4 Exercising at Home
- 3.5 The National Institutes of Health
- 3.6 Medical-Identification Jewelry
- 3.7 Being Prepared
- 3.8 Handicapped Stickers
- 3.9 Staying Healthy



### 3.1 Coping with Your Diagnosis

When you were diagnosed, you probably had countless feelings accompanying the news. You might have experienced feelings of anger, denial, shock, grief, helplessness, confusion, despair, sadness, and fear. None of these feelings are uncommon for women diagnosed with LAM, and many women experience one or all of them during various stages of the disease. These emotions are all normal responses and are all part of the grieving process. What!? No one died, so you're not grieving? Grief isn't just related to death; it can be the result of any loss—including the loss of the lifestyle you once enjoyed. Understand that you'll probably grieve some because of some of the changes you may face with LAM. Many changes may be far in the future for you, so it's best not to dwell on your disease.

You may experience another common reaction to your diagnosis: relief. If, over a long period of time, you, like many patients, have experienced symptoms like shortness of breath or a lung collapse without your doctor finding a reason for your problems, you'll probably be relieved to finally have a name to tack onto your health issues. You'll feel great knowing you didn't imagine all these problems.

As with any chronic illness, your family and friends will want to provide emotional support for you. But you need to understand that your closest supporters may not feel any more comfortable in their new roles than you do. Your support people will need you to tell them how they can help you so that they don't feel helpless and powerless in their relationships with you. Just like you, your family and friends will experience a flood of emotions and will have many questions regarding your diagnosis. Do your best to keep an open line of communication with them and keep them informed. Such actions will ensure that your relationships remain as healthy as possible. Being treated differently is one of the greatest fears for most LAM patients. If you don't want to be treated differently, tell them that.

A diagnosis of LAM is difficult all by itself, but a lack of communication can lead to marital stress or parental concerns. Financial worries, insurance coverage, and employment issues can also trigger additional worries and anxieties at this time. To help deal with these stresses you may want to consider complementary therapies to nurture your body, mind, and spirit. Exercise, acupuncture, massage therapy, yoga, and reflexology are just some ways to relax and to help your coping skills. If the stresses accompanying your diagnosis of LAM become overwhelming, don't hesitate to seek emotional support from a friend or professional help from a psychologist, a counselor, or a member of the clergy. Many LAM patients have done just that. Learning how to manage stress, anxiety and depression is necessary in all phases of life, but it's even more essential in maintaining an optimum level of health now that you have a chronic disease.

You've probably just gone through an exhaustive battery of tests that determined your diagnosis. Unless you're in a critical state, you don't have to decide on a treatment plan immediately. Take some time to absorb the fact that you have LAM. Be patient with yourself.

Your doctors and The LAM Foundation probably sent you a barrage of information. Read the material you received slowly. Visit The LAM Foundation's website (<https://www.thelamfoundation.org>), read the *Currents e-newsletters* and *Journeys* newsletters. Make contact with other LAM patients through the LAM Liaison Program or the Lammies Facebook page (see the Patient Services section to learn more). The LAM Foundation

offers these and other programs to support you and your family. Don't hesitate to call the staff at the Foundation to ask for their assistance.

When you're comfortable with your diagnosis, consider attending the annual LAM conference, LAMposium. During this meeting, LAM patients travel from all over the world to share the camaraderie during this wonderful weekend. You will be able to attend informational sessions to learn more about LAM. Medical professionals, who work with LAM and who are themselves attending concurrent clinical and scientific sessions, give presentations to women with LAM and their families, and they are available to answer your questions. It's an opportunity to spend time with other women with who know what you're feeling.

Finally, when the dust settles, you might want to visit the National Institutes of Health (NIH) to find out more about your disease and to meet other women with LAM. You'll find that education is the key to empowering yourself. You'll realize that you have a choice: to live productively with LAM or to allow it to control your life.

### **3.2 Pulmonary Rehabilitation**

When you got the news that you had LAM, one of the first questions you probably wanted answered was how to stay healthy. Unfortunately, there is no simple answer to that question. But there is one thing—and one thing only—that appears to help almost every single LAM patient: exercise.

E-X-E-R-C-I-S-E. One, two, three, four, five, six, seven, eight. Eight letters. You must forget your conviction that exercise is a four-letter word. Now don't go saying that you're having a tough enough time breathing without adding exercise to your life. Almost all women with LAM have difficulty breathing. But each woman who exercises—no matter how little she exercises—feels better (maybe not immediately, but in the long run) for doing so. Doctors have seen great results from women who exercise.

Many women with LAM exercise. Some have developed good routines of running, walking, weight training, or yoga, while others participate in recreational sports such as tennis, swimming, or biking. You can't do any of those things? Well, maybe not. But, you can get moving. And, more importantly, you *should* get moving.

If you're not already active, the best way to get started is in a pulmonary-rehabilitation program. Ask your doctor to refer you to a pulmonary-rehab center that is near you and that fits your needs. And don't forget to remind your doctor what kind of insurance you have.

At pulmonary rehab, exercise physiologists or other trained personnel supervise your workouts. This supervision is very important because your oxygen-saturation level (the level of oxygen in the blood, called an "O<sub>2</sub> sat") must be closely monitored to insure that the exercise is helping—not hurting—you. The people at pulmonary rehab are specially trained to help you improve your level of fitness. They'll devise a workout for you, and they'll monitor you every step of the way.

#### **What Can Pulmonary Rehab Do for You?**

Rehab isn't just a gym where you exercise. Think of it as a full mind-and-body tune up. Are you housebound? Are you having difficulty performing duties at work, or is running a

household becoming more than you can deal with? Are you anxious when you drive? Believe it or not, pulmonary rehab can help you address all of these problems. Depressed? Rehab can even help you psychologically. For example, if you go to rehab regularly, you'll develop relationships with your therapists and the people you work out with. You may even become friends with some of them. These added social interactions can decrease your level of isolation if your disease has progressed to the point where you don't go out much. Increasing social interactions can help lower your level of depression.

The staff working in a rehab center is specially trained to teach you how to move on with your life, and becoming more fit is just the beginning. LAM affects your entire body, not just your lungs. But, as you become more fit, you'll learn your body's capabilities and you'll become more confident. You'll feel more comfortable in your ability to live as you choose to.

If your lung disease has progressed far enough for you to be evaluated and listed for a lung transplant, you'll discover that most lung-transplantation teams require your attendance at a pulmonary-rehabilitation program as a part of your preparation for transplant surgery. Transplant doctors want you to be as strong and healthy as possible before you receive your new lungs. Not only will you recover more easily from the surgery, but you'll also enjoy an improved quality of life.

### **The Evaluation**

Before you begin your workout at rehab, you'll go through an evaluation process. In an introductory interview, you'll be asked to describe what impact LAM has had on your lifestyle and what aspects you wish could be improved to increase your overall sense of physical and mental well-being. This is a time to be honest. You aren't going to rehab to be judged. The people who work there want to help you. Do you get upset more often than you used to? Are you having trouble with the stairs at work? Can't vacuum anymore without becoming extremely short of breath (SOB)? By talking openly with the respiratory therapists, together you'll design a personalized program to help you overcome some of these problems. The staff can teach you breathing techniques to calm you or to help you climb stairs more easily. They may even put you in touch with an organization that will send someone out to vacuum and clean for a minimal fee. Rehab, in general, is a terrific resource. Plus, most rehab visits last only about an hour.

As with many medical interviews, you'll be asked about your medical and surgical history and that of your family. The interviewer may also ask whether you smoke or have in the past or whether you have a history of drug and/or chemical use or abuse. The greatest focus, obviously, will be on your respiratory problems and *your perception* of how these problems affect you. Now is the time to talk about your respiratory symptoms like shortness of breath (dyspnea), fatigue, cough, chest pains, wheezing, and lung collapses (pneumothoraces).

In the physical assessment, you'll probably go through a very in-depth, intensive diagnostic workup that may even include an ECG and/or a stress test and oxygen monitoring. Many patients have already experienced such tests at the National Institutes of Health (NIH) or at the request of their personal physicians, so rehabilitation centers often accept your results from these tests if the test scores are recent. Some centers will perform a less intense physical assessment or will just start you on a low-intensity program, monitoring you and increasing your routine as your fitness progresses.

When you go, you'll probably be weighed, and the person checking you in will ask about your medications, your general health, and how you feel that day. Actually, every time you go to rehab you'll be asked about your general health that day and your level of breathing.

Before you start any exercises, the technician or nurse will take your vital signs: your blood pressure, heart and respiratory rates. He or she will also check your oxygen-saturation level using a pulse oximeter (a small device that clips onto your fingertip) and will listen to your heart and lungs, especially if you complain—or have a history—of fluid retention.

### **The Six-Minute Walk**

Eventually, whether it's at the NIH, at a doctor's office, or at a rehab center, all LAM patients perform the "six-minute walk." What is it? It's exactly what it says it is. You walk for six minutes and are assessed. Generally, a respiratory therapist will administer the test. First, he or she will clip a pulse oximeter on your finger or stick a pulse sensor to your forehead. Then, you'll carry the portable oximeter, a very light, little machine, and walk for six minutes. While you're walking, the therapist will time you, note your saturations and pulse measurements at minute intervals, and monitor your exertion levels as you describe them, according to a scale of perceived exertion (to be explained later in this chapter). The point of the test is for you to walk as quickly as possible to see if, or how low, you desaturate, that is, if your oxygen-saturation level drops. The test measures your heart rate as well. When you have finished walking, you are monitored for a few more minutes to see how long it takes your body to recover from six minutes of walking. The results of the test may indicate that you need oxygen for certain activities or that your disease has progressed.

### **The Workout**

After you've been checked over and have answered a ton of questions, you're ready to exercise. Your therapist will ask you to perform several different exercises. You may walk a few minutes on a treadmill, do a few minutes on a stationary bicycle, a few minutes on the arm ergometer, lift some light weights, or work on your flexibility. When you begin, you may only be able to do a minute or two at each station. But as you become stronger, you will be able to do more. Your goals should be to do thirty minutes of exercise on either the bike or the treadmill and to reach your target heart rate, which your therapist will help you calculate. In about four to five weeks, you'll begin to notice changes in your body and in your ability to function. You'll start to feel so much better, and you should be able to do more than you could have before you began exercising.

## The Scale of Perceived Exertion

When you're exercising at rehab, the staff will occasionally ask you to rate your level of breathlessness and/or exertion according to a scale of perceived exertion\* similar to the one presented here.

0	Nothing at all
.5	Very, very slight (just noticeable)
1	Very slight
2	Slight
3	Moderate
4	Somewhat severe
5	Severe
6	
7	Very severe
8	
9	Very, very severe (almost maximal)
10	Maximal

Your therapist wants to see how your perception of breathlessness and exertion corresponds to your oxygen-saturation levels and your heart rate. Such data helps to monitor you and your progress.

## Education

Besides physical conditioning, many rehab centers offer educational sessions on a variety of topics to teach you how to live better. Topics may include information on pulmonary diseases, what medications can aid breathing and how to take them efficiently, nutritional sessions geared for people who have trouble breathing, and stress-management techniques. You'll also learn pursed-lip and diaphragm breathing techniques and other tips to make your daily activities, and your life, easier. To get the most from your rehab experience, you should attend these seminars even if they are not required.

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\* Gunnar Borg, PhD, is the developer of the Perceived Exertion Scale. See the page, following this chapter, on the Borg Ratings of Perceived Exertions Scale for a more detailed description.

## Oxygen

The volume of your supplemental oxygen flow is measured in liters per minute (LPM). If you are already using supplemental oxygen, the people at rehab will help you learn what liter flow you need for different kinds of activities. You'll learn what liter flow you need to perform all the tasks you face every day. For example, when you're vacuuming, you might require four or five LPM, but when you're watching TV, you can turn the flow down to two liters.

If you have not used oxygen before, the rehab staff will advise you if they feel oxygen would help you exercise more safely and productively. At pulmonary rehab, supplemental oxygen is available to those who need it, and your oxygen saturations are monitored so that you're always getting sufficient oxygen. If your oxygen saturation drops while you're on the treadmill, try to continue at your assigned speed, but increase your oxygen flow—this will maximize the effectiveness of your efforts. And, if you exercise at a pulmonary rehabilitation center, while you're there you can generally use their oxygen at no cost to you. What a bonus!

## Cost

Many insurance companies pay for a set number of visits for pulmonary rehabilitation. Your doctor will probably ask you to go there two or three times a week and you may find that your covered visits are soon used up. Many centers, though, offer inexpensive programs to keep you coming to rehab. Often, the cost is less than five dollars a visit, and it's worth every penny.

## Summary

Remember: Exercise is not just good for you; it is *necessary* for you. It's a matter of quid pro quo: that is, the more you work at something, the more you'll get out of it. So, ladies, if you're exercising already, three cheers for you. And if you're not, get started as soon as you talk to your doctor.

## Some Possible Benefits of Exercise

- Lowers your resting blood pressure
- Lowers your total cholesterol
- Increases your HDL
- Controls blood-glucose levels; decreases insulin use
- Decreases anxiety and depression
- Improves functional strength, stamina, balance, and coordination
- Improves self-esteem
- Prevents premature aging
- Improves bone density
- Increases psychological and social wellness
- Improves sexual relations
- Decreases your body fat and weight

## Goals of Pulmonary Rehabilitation

- To control or alleviate respiratory symptoms and complications
- To increase exercise tolerance
- To decrease anxiety and depression
- To teach how to carry out activities of daily living
- To improve quality of life
- To promote independence and self-reliance
- To reduce hospitalizations
- To promote social and recreational interactions
- To encourage employment options
- To ensure cardiac well-being through adequate O<sub>2</sub> levels

When you join a pulmonary rehabilitation program, you need to understand one major point: You are the person going to rehab. The staff there will help you and guide you, but ultimately, rehab is only as successful as the effort you put into it. Go with a positive attitude—not because you have been forced to go. Go because you want to be healthy. Go to make your life the best it can be.

### 3.3 Borg Rating of Perceived Exertion Scale

The Borg Rating of Perceived Exertion Scale (Borg RPE scale, © Gunnar Borg, 1970, 1985, 1994, 1998) is used in varied forms (such as the one given in the Pulmonary Rehabilitation section) by many rehabilitation centers. It is given in full here with some notes to further explain the stages.

#### The Borg Rating of Perceived Exertion Scale

6	No exertion at all
7	
7.5	Extremely light
8	
9	Very light
10	
11	Light
12	
13	Somewhat hard
14	
15	Hard (heavy)
16	
17	Very hard
18	
19	Extremely hard
20	Maximal exertion

9 is "very light" exercise. For a healthy person, it is like walking slowly at his or her own pace for some minutes.

13 is "somewhat hard" exercise, but it still feels OK to continue.

17 is "very hard" or very strenuous. Healthy people can still go on, but they really have to push themselves. It feels very heavy, and the person is very tired.

19 is "extremely hard" or an extremely strenuous exercise level. For most people this is the most strenuous exercise they have ever experienced.

### 3.4 Exercising at Home

Not all women join a pulmonary rehabilitation program when they're first diagnosed with LAM. Many walk daily, either outdoors or on a treadmill. Others bike or play tennis. But if you're not in a monitored class, here are some tips for sticking with an exercise program:

1. If you miss a few days—or weeks—of working out, don't feel guilty. Acknowledge that you skipped your workouts and just start exercising again. Don't belittle yourself. What's passed is past; the rest of your life begins today.
2. Mark your calendar. Plan your workouts the way you plan the rest of your life. Write them down. Set aside at least 30 minutes a day, three times a week.
3. Remember the buddy system? If you've been trying to exercise alone, and failing, get a workout partner or join a class. Skipping a workout when you exercise alone is really easy. But if you're meeting someone, it's harder to cancel. And remember: showing up is half the battle.
4. Trick yourself into a five-minute workout. If 30 minutes are too daunting, and you hear yourself saying that you don't have the time, just work out for five minutes. Maybe ten. Once you get going, chances are you'll want to continue.

Take it easy. One of the main reasons people quit exercising is because they try to do too much at the beginning. They push themselves too hard. The fix is simple: SLOW DOWN and STICK WITH IT!

**As always:**

**CHECK WITH YOUR DOCTOR BEFORE STARTING ANY EXERCISE PROGRAM.**

### 3.5 The National Institutes of Health

The National Institutes of Health (NIH), located in Bethesda, Maryland (USA), is the biomedical research agency of the federal government. LAM research began there in 1996. You may hear people refer to the NIH as the NHLBI, the National Heart, Lung, and Blood Institute. The NHLBI is one of about twenty-two Institutes that make up the NIH. The NHLBI is conducting a LAM protocol (study). This LAM research program has been set up to study the development and course of LAM at the cellular and molecular levels.

Many LAM patients participate in this research protocol and travel to Bethesda every six months for evaluation. Upon arrival, patients are admitted to the Clinical Center to undergo different tests\* which may include, but are not limited to, lab work, chest x-rays, computerized-tomography scans (CTs), magnetic-resonance imaging (MRIs), pulmonary-function tests (PFTs), electrocardiograms (EKGs), bone-density tests, exercise tests, and abdominal ultrasounds. Patients may decline any test that they feel uncomfortable about but most women participate fully knowing that their wholehearted involvement may help find a cure for LAM. The results of all these tests are compiled into a database that is a cornerstone for research and treatment.

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\* These tests are free for all LAM patients in the protocol. The clinic stay, meals, and sometimes transportation, everything but the souvenir T-shirt, are free too. Foreign patients, however, must pay their own transportation to the States, but then they'll receive all of the free benefits of a NIH visit.

The protocol is set up so that you'll have one long visit and one short visit each year. During your initial visit and on your annual long visit, you'll have the majority of your tests, and you may stay for four or five days. The short visit may last only a couple of days and usually consists of lab tests, PFTs, EKGs, and a chest x-ray. You'll receive notification of your next NIH visit about two months ahead of time. Don't hesitate to call the NIH to negotiate different dates to fit your schedule. No one expects you to miss your child's birthday or your parents' anniversary party to check into the hospital. But call the NIH well ahead of time if you need to change your admission date. The staff needs advance notice to reschedule your tests, and they won't be able to accommodate you if you wait until the last minute to call. If you have a busy schedule at home, the staff at NIH will squeeze your tests into as small a time as possible so you can get back to your family and/or your job.

When you were first diagnosed, you probably had more tests than you ever thought existed. Why in the world would you ever want to go to the NIH to have even more tests? Obviously, most women don't go because they like the tests. Many went originally because their own doctors knew so little about LAM. These women had zillions of questions that they needed answered, and the NIH is considered the pre-eminent source of information about LAM. Now, many women continue the protocol because they enjoy it. You might enjoy it too. The NIH is a safe haven where you can go to ask any questions you have; the staff is made up of people who specialize in LAM. It's a meeting place to talk with other women with LAM who understand exactly how you feel, and it's a shelter to express your fears. And the NIH can be a place to have a good time.

Have a good time in a hospital? Yes. The entire staff—technicians, nurses, nurse practitioners, and doctors, are incredibly friendly and wonderfully supportive. They understand that you are helping them with their research, and they treat you as a welcome and respected guest. Doctors, nurse practitioners, and nurses all give you inordinate amounts of time, time you could never spend with your doctors at home. But when you're done with your tests for the day, you have the opportunity to get out and meet the other LAM patients who are there in the wing with you. Many of these women are there to see old friends, friends they've met at NIH. They also want to meet you!

Generally, several other LAM patients will be at the NIH with you. These women will be in different stages of LAM. You may meet newly diagnosed women, women who have had a lung transplant and women who are stable and who've just been going about their daily routines, not allowing LAM to rule their lives. You can sit around talking to these women, or you can join them in activities offered by the NIH. You are encouraged to wear your own most comfortable clothes and to use your free time in whatever way you choose. If you're a crafter, the fourteenth floor has a well-stocked craft room. For dog lovers, dogs occasionally visit to give you some extra TLC. Even Tai Chi and relaxation courses are offered. Just hanging out or exploring the grounds can also be an adventure. And, if you have large blocks of time, you can sign out and take the subway into Washington, DC, and explore our nation's capital, or you can just go into Bethesda for a nice dinner.

It's natural to be nervous about your first visit to the NIH. To calm your fears, feel free to bring your husband, your partner, another family member, or a friend. Although your companion cannot stay in your room overnight, he or she can find lodging in the Bethesda area to suit any budget. After their initial visits, many women choose to return to the NIH alone. They find it more relaxing to do whatever they want to without taking another's feelings and

needs into account. For some women with hectic daily schedules, just relaxing in bed all day reading or watching TV may benefit them more than sightseeing. Knowing they don't have to cook, clean, or do laundry for a few days is a blessing for others. You will even have time to pamper yourself.

Are you concerned about the cost of going to the NIH? Don't be. The NIH pays for transportation for LAM patients who live in the United States. (Patients from other countries who are willing to pay their own transportation costs and who can participate in the study are also welcome to join the protocol. The NIH will pay travel expenses within the US or Canada.) After your appointment is scheduled, you can call the NIH travel agent to book a flight or other mode of transportation. You may also travel by train, bus or in your personal car, whatever you like. If you travel by car, you will be reimbursed for mileage. If you fly, free shuttles from the airport are available. For those who are on oxygen, an NIH driver, with oxygen for you, will meet you at the airport or train station to take you directly to the NIH.

At the end of your stay, you will have an exit interview with one of the staff physicians and the nurse practitioner assigned to you during your stay. These two professionals will give you a summary of your test results and will give you any recommendations they have. Then they'll invite you to ask questions and will take all the time necessary to answer them. When you leave the NIH, you'll have a copy of most of your test results: e.g., lung-function tests, lab work for blood and urine, and exercise-test results. Frequently, these test results may substitute for testing that you would have done at your home hospital, and you won't need to pay for tests done at NIH. A copy of all of these results, along with a summarizing cover letter, will be sent to your doctor at home. You are encouraged to meet with your personal physician to go over the results and, based on the NIH recommendations, to plan any changes in your treatment plan and/or lifestyle.

If you're interested in participating in the NIH research protocol, enrolling is easy. Just call the nurse in charge of the study at 1-877-644-5864 (choose selection three on the menu) to introduce yourself and to ask what you need to do to sign up. You'll be glad you did.

You can find out more about the National Institutes of Health by checking out the NIH website at <http://www.nih.gov>. To see if you qualify for the LAM protocol, check out <http://clinicaltrials.gov>, and, at the prompt for the disease, type in LAM or lymphangioliomyomatosis.

### **3.6 Medical-Identification Jewelry**

When a person has a medical condition like LAM that sometimes requires emergency care, wearing medical-identification jewelry is a good thing. Medical-identification jewelry is generally sold in a choice of bracelets or necklaces with a charm attached that bears the wearer's pertinent medical information.

There are several sources for medical jewelry. Drug stores sell generic pieces inscribed with common diseases and conditions like diabetes, asthma, and drug allergies. You can also select a piece of jewelry that just says, "See Wallet Card." All of these bracelets and necklaces come with information cards to carry in your wallet or purse. These cards can be filled in with extra data such as your name, address, and phone, your doctor's name and phone and perhaps your

current prescriptions. Pharmacies usually carry application forms so that you can order jewelry with special engraving for rare diseases or extra medical information. If you can't find these forms, ask your pharmacist, call the reference department at your local library and ask for help in locating a company, or search for "medical jewelry" on the Internet.

If you're in the LAM protocol at NIH, you've probably received one of these generic bracelets on your first visit, but the staff there will urge you either to keep your wallet card handy at all times or to have a more specialized bracelet made. You can choose from a wide range of styles and prices if you decide to get a custom-made bracelet or necklace.

There are also specialty firms that deal with such jewelry. One such firm is MedicAlert®. You can contact them online at <http://medicalert.com> or call them at 1-888-633-4298 (toll-free) for their brochure. MedicAlert® jewelry is sold in conjunction with an emergency medical-notification service that is on call twenty-four hours a day. You buy the service (around \$30-40 the first year and \$15 each subsequent year) along with your jewelry, and the company keeps your medical information on file in case of an emergency. The company's toll-free phone number is engraved on the charm along with your pertinent medical information.

Along with the notification service, MedicAlert® will confidentially store an advance directive for you so that it's available twenty-four hours a day, whenever you need it in an emergency. The fee for this service is about \$30 per year.

Lauren's Hope, another company, specializes in beaded medical-identification bracelets. Although many people prefer a bracelet that isn't as institutional-looking as some identification bracelets, make sure that your bracelet still retains the medical "look" to get the attention of emergency crews so they can find your necessary medical information. You can visit the Lauren's Hope website at <http://www.laurenshope.com> or call 1-800-360-8680 for help with ordering, engraving, or just to ask a question.

If you'd like to see some pieces of jewelry before you buy, look for stores in your area, especially in malls, that sell and engrave items like pens, cups, and other gifts. The medical-identification jewelry sold in these stores is often quite expensive, but buying your jewelry there allows you to see what you're getting and to consult with the sales staff about what information will fit well on any given item.

## **What to Engrave on Your Jewelry**

Some LAM patients have “lymphangioleiomyomatosis” spelled out and include a phrase like “diffuse lung disease” because LAM is so rare. You can also add “possible lung collapse” or “potential for lung collapse” if you’re prone to pneumothoraces. You can even add that you’ve had a lung pleurodised to guard against someone puncturing it during a medical procedure. Use caution with your word choices because not all emergency medical personnel are familiar with all of the words that have become part of your working vocabulary. You may consider saying “lung collapse” instead of “pneuomothorax,” and “lung adhered to chest wall” instead of “a pleurodised lung.” Finally, adding your physician’s name and number to your jewelry is also helpful.

If you’ve received a transplant, most centers require that you wear medical-identification jewelry, detailing your unique medical condition. Information can include your name, what organ was transplanted, the fact that you are on immunosuppressive drugs, and the local and toll-free phone numbers of the transplant office. Drug allergies should also be noted.

### **Tips**

Many people find that these identification bracelets and necklaces are not at all intrusive or bothersome, and they wear them all the time. Others wear them only when they go out. Some women carry short medical histories, current medications, drug allergies, and emergency names and phone numbers with them in their purses. The bracelet, the necklace, the card, and the short medical history are all good ideas, so choose whichever options best fill your needs. Think about the amount of traveling you do and the number of times you might be in a situation when you need to rely on the help of strangers. Your lifestyle should help you determine how you identify yourself as the special person (LAM patient) you are.

## **3.7 Being Prepared**

Having a chronic medical problem can complicate your life in ways you never anticipated when you were first diagnosed. For example, if you need to go to the emergency room (ER) and are so breathless you can’t speak, the ER doctors still need to know something about you, your general condition, and your medications. While this is true to a degree for everyone, your medical history is probably a bit more complicated than that of the average person.

Travel can also cause problems if you forget to pack your medications, run out of oxygen or rent a faulty concentrator.

To help you cope with trips to the ER and to make your travel more enjoyable, we’ve compiled some tips to facilitate service from people who may not know you very well. We’ve also included several pages that can be removed and copied, so you can fill them out to keep your vital information on hand. Start a personal medical packet to take with you on trips to the ER, on vacation, and when you see any doctor for the first time.

**For the Emergency Room:**

- Have your medical information placed where your family and/or friends can locate it, and be sure they know where it is.
- Keep an additional copy of this information in your car.
- Put an “Emergency Medicine Quick Facts for Lymphangiomyomatosis” card in your wallet where it can easily be seen or in an iWallet on the back of your cell phone.
- Bring your cell phone and a charger with you.
- Bring along a headset or a good book.
- Take your health-insurance card(s).
- Wear a medical –identification bracelet or necklace.

**For Travel:**

- All of the above tips for visits to the emergency room apply plus:
- Give another copy of your medical information to a friend back home in case you lose your copy.
- Pack extra medications while traveling in case of delays or lost luggage.
- Keep all medications in your carry-on bag. Never check them with your baggage.
- Carry a doctor’s prescription for supplemental oxygen.
- Have the names and numbers of your regular oxygen supplier and your travel supplier with you.

**General Information**

You should know your height and weight, your normal blood pressure and temperature and your blood type. Jot down recent changes in your medications, such as dose changes or the deletion or addition of certain medications. Be ready to tell the doctor when you had your last flu shot and your last pneumovax (Pneumococcal vaccine) and whether you had any medical treatments, lab tests or procedures such as dental work. Other items that are handy to have are copies of your CT scans (NIH is now sending you home with a CD with copies of your latest scans on it.) and/or x-rays and PFTs if you have them. If you’ve had a transplant, spirometry records are useful for any doctor in the ER.

Finally, if you have an advance directive, notify the medical staff of its existence, give them a copy and tell them where the original can be found.

All of this preparation probably seems excessive and you should understand that not all of these items are needed at all times. Some may never be needed. But planning ahead and gathering this information may be just what a doctor will need to treat you or what you need while traveling. And having this information ready may also put your mind at rest knowing that you’re well prepared for any emergency.

# Personal Information

Name: \_\_\_\_\_

Address: \_\_\_\_\_

Home Telephone: \_\_\_\_\_

Office Telephone: \_\_\_\_\_

Date of Birth: \_\_\_\_\_

Social Security Number: \_\_\_\_\_

Contact person: \_\_\_\_\_

Relationship: \_\_\_\_\_ Telephone: \_\_\_\_\_

Telephone: \_\_\_\_\_

Medic-alert company: \_\_\_\_\_ Telephone: \_\_\_\_\_

# Medical Insurance Information

Insurance Company: \_\_\_\_\_

Address: \_\_\_\_\_

ID Number: \_\_\_\_\_ Telephone: \_\_\_\_\_

Group Number: \_\_\_\_\_ Fax: \_\_\_\_\_

**Tip:** Keep a copy of your insurance card(s) handy.

# Physician Information

Include all physicians and your transplant coordinator.

Name: \_\_\_\_\_ Specialty: \_\_\_\_\_

Address: \_\_\_\_\_ Telephone: \_\_\_\_\_

\_\_\_\_\_ Fax: \_\_\_\_\_

Name: \_\_\_\_\_ Specialty: \_\_\_\_\_

Address: \_\_\_\_\_ Telephone: \_\_\_\_\_

\_\_\_\_\_ Fax: \_\_\_\_\_

Name: \_\_\_\_\_ Specialty: \_\_\_\_\_

Address: \_\_\_\_\_ Telephone: \_\_\_\_\_

\_\_\_\_\_ Fax: \_\_\_\_\_

Name: \_\_\_\_\_ Specialty: \_\_\_\_\_

Address: \_\_\_\_\_ Telephone: \_\_\_\_\_

\_\_\_\_\_ Fax: \_\_\_\_\_

Name: \_\_\_\_\_ Specialty: \_\_\_\_\_

Address: \_\_\_\_\_ Telephone: \_\_\_\_\_

\_\_\_\_\_ Fax: \_\_\_\_\_

# Medications

## Prescriptions

Drug	Strength	Dose	Time Taken	Reason for Taking

## Over-the-counter (OTC) Medications

Drug	Strength	Dose	Time Taken	Reason for Taking

## Vitamins and Herbal Supplements

Drug	Strength	Dose	Time Taken	Reason for Taking

## Supplemental Oxygen

When: \_\_\_\_\_ Liter flow: \_\_\_\_\_

When: \_\_\_\_\_ Liter flow: \_\_\_\_\_

When: \_\_\_\_\_ Liter flow: \_\_\_\_\_

Oxygen  
Supplier: \_\_\_\_\_ Telephone: \_\_\_\_\_

## Pharmacy

Name: \_\_\_\_\_ Telephone: \_\_\_\_\_

Address: \_\_\_\_\_ Fax: \_\_\_\_\_

\_\_\_\_\_

## Allergies

### If you have no allergies,

use NKDA (no known drug allergy) or NKFA (no known food allergy).

Drug: \_\_\_\_\_ Reaction: \_\_\_\_\_

Drug: \_\_\_\_\_ Reaction: \_\_\_\_\_

Drug: \_\_\_\_\_ Reaction: \_\_\_\_\_

Drug: \_\_\_\_\_ Reaction: \_\_\_\_\_

Food: \_\_\_\_\_ Reaction: \_\_\_\_\_

Food: \_\_\_\_\_ Reaction: \_\_\_\_\_

Food: \_\_\_\_\_ Reaction: \_\_\_\_\_

Food: \_\_\_\_\_ Reaction: \_\_\_\_\_

Other: \_\_\_\_\_ Reaction: \_\_\_\_\_

Other: \_\_\_\_\_ Reaction: \_\_\_\_\_

Other: \_\_\_\_\_ Reaction: \_\_\_\_\_

Other: \_\_\_\_\_ Reaction: \_\_\_\_\_

(Other allergies may include an allergy to latex or to certain environmental factors.)



## **Pneumothoraces**

When: \_\_\_\_\_ Side: \_\_\_\_\_

Treatment: \_\_\_\_\_

## **Chylothoraces**

When: \_\_\_\_\_ Side: \_\_\_\_\_

Treatment: \_\_\_\_\_

**Hospitalizations**

When: \_\_\_\_\_ Where: \_\_\_\_\_

Why: \_\_\_\_\_

\_\_\_\_\_

**Surgeries**

Surgery: \_\_\_\_\_ Where: \_\_\_\_\_

When: \_\_\_\_\_

### 3.8 Handicapped Stickers

Getting tired of walking a long way with your oxygen tank just to get from your car to the store? What if you're not on supplemental oxygen, but you're still getting tired just trying to shop? Would a handicapped sticker help you so you could park closer to your destination?

If you answered "yes" to any of these questions, you could be eligible for a handicapped sticker for your car. Technically, this sticker or hanging card is called an ADA (Americans with Disabilities Act) placard.

Don't be shy about asking your doctor if you might be ready for this kind of help. If it's hard for you to perform your daily activities and if parking closer would help, you may very well have the requisite PFT numbers to qualify. A quick online check for the State of Michigan ([http://www.michigan.gov/documents/bfs-108\\_16249\\_7.pdf](http://www.michigan.gov/documents/bfs-108_16249_7.pdf)) revealed the following criteria (these should be basically the same in other states) on its application for an ADA placard:

*Patient has lung disease from which the forced expiratory volume for one second, when measured by spirometry, is less than one liter, or from which the arterial oxygen tension is less than 60 mg/hg of room air at rest.*

or

*Patient has a persistent reliance upon an oxygen source other than ordinary air.*

Many women with LAM meet these requirements and you may be one of them. The local office that handles car registration and license plates will guide you through the process. It usually involves bringing documentation from your doctor, and after your application is processed, you'll be issued a placard. There will probably be a small fee.

Once you get your ADA placard, you can legally park in a handicapped spot. If you park in one and you aren't on supplemental oxygen, someone might come up to you and give you a hard time about your being too healthy to park there. Don't get angry or defensive. Understand that not all disabilities are visible ones. Try to turn this negative event into a positive one. Keep a handful of LAM brochures in your car for occasions like this. Give one to the insensitive busybody and raise awareness of LAM at the same time!

One more bit of advice: ADA placards are very attractive to less-than-honest people. Keep your car locked or put the placard in your glove compartment or some other out of sight location, or you may return to find that it has been stolen! Replacement placards are available at the same motor vehicle registration office, but a little caution will save you the trouble of getting a new one.

### 3.9 Staying Healthy

OK, so you have LAM. What are you going to do about it? Obviously, there isn't a lot you can do because little is known about the disease, but you *can* try to stay as healthy as possible. Here are some things that you can and should do.

One: Don't smoke. If you smoke now, you need to quit. Smoking anything—from cigarettes to marijuana—will only make your sick lungs sicker. You should also avoid secondhand smoke. Remember, you have every right to a smoke-free home or office.

Two: Watch your diet. Many women with LAM probably don't need a special diet, but everyone needs to eat healthy meals, including five or more servings of fruit and vegetables a day. Eat lean protein, add calcium-rich foods to your diet to help maintain your bone density (most LAM patients do suffer from varying degrees of osteoporosis and/or osteopenia), and limit your intake of sugars and fats.

Since estrogen is suspected of playing a major role in this disease, many doctors recommend that LAM patients avoid soy products because they contain large amounts of phyto-estrogens. Phyto-estrogens are estrogens that occur naturally in plants, but these estrogens may also play a role in LAM. Researchers don't know yet if plant estrogens are harmful to women with LAM, but eating these products—and foods of all types—in moderation would be a good idea.

If you have trouble breathing after meals, try eating five or six smaller meals rather than three big ones. Sometimes if your stomach is too full, your diaphragm doesn't have room to expand, and you feel short of breath.

For women who have difficulty with chyle (see the section on Chylothorax and the related problems) following a low-fat diet can be essential in keeping chyle production to a minimum. If you are experiencing this problem, consult your doctor or the staff at NIH to learn more about chyle and how to control it. If you participate in the protocol at the National Institutes of Health, you can request an appointment with a dietician to teach you how to minimize the problems you experience.

Another incentive to eat well and in moderation is that women who are transplant candidates need to have their weight within a normal range. If you're approaching transplant time, work to lose or gain pounds sensibly to get your weight into the target range.

Three: Exercise. Exercise may be the most important thing you can do to remain at your optimum level of health. Stay as active as possible. Women with LAM know how difficult those simple words are but those who exercise tend to have fewer problems. No one expects you to go out and run a marathon but if you start slowly and gradually build yourself up, you'll have more endurance to perform even the simplest household tasks. Exercise will maximize your body's ability to absorb oxygen, increase your energy and make you a better candidate for transplant. To get started on a safe and effective exercise program, consult your doctor to learn about pulmonary rehabilitation.

Four: Get plenty of sleep. Sufficient sleep is essential to your health. Your pulmonologist may recommend that you use supplemental oxygen when you sleep. If you need it, you should use it. Nighttime O<sub>2</sub> can make a big difference in how you feel during the day.

Five: Avoid stress. Naturally, that's easier said than done, but you should find a way to lower your stress level whether it's sitting and having a second cup of tea, taking a yoga class, or going to a movie. Try to manage your stress in a healthy manner.

Six: Schedule regular appointments with your doctor. Because you have LAM, little problems can grow into big ones in no time at all. Regular medical (and dental) appointments can help you and your doctor ward off many potential problems and keep current problems from growing.

Seven: Take your prescription medications as indicated. Medicines work best when they're taken as directed. Try not to skip pills or take extra ones and don't stop antibiotics until the dose is complete unless your doctor tells you to stop.

Eight: Get a flu (influenza) shot. Many doctors recommend flu shots for all of their patients who have chronic health conditions—especially heart and respiratory problems. As a LAM patient, you should consider getting a yearly flu shot. It's especially important to get the vaccine if you work in an institutional setting such as a school or health-care facility where you are exposed to many people. You might also encourage those you come into regular, close contact to do the same.

The flu vaccine is given to protect people from contracting influenza, a viral illness that affects the respiratory tract. In the US, the shots are usually given in late October or early November because the flu season generally begins in late November and lasts until April. Your body needs ample time (about two weeks) to build immunity, so check with your doctor to see when you should have the vaccine. Ideally, you want the vaccine's effectiveness to last throughout the worst of the flu season. But don't hesitate to get a flu shot at any time during flu season!

For many healthy people, the illness often lasts only 3 to 7 days, but those less healthy may have more severe cases and even require hospitalization. And don't forget: people can die from the flu.

Because different strains of influenza exist, a flu shot is required every year. The yearly vaccines are developed to protect people from the three major strains that are expected to be the most prominent each year. You cannot get the flu from the flu vaccine, but you may experience tenderness at the injection site, minor achiness, or a low-grade fever after you receive the injection. Check with your doctor to see if you should be getting a yearly flu shot.

Nine: Get a Pneumococcal vaccine (pneumovax). Many doctors are now recommending that patients with chronic heart and respiratory conditions receive a pneumovax. Generally, this is a single-dose, one-time injection to prevent people from developing pneumonia. Even though one dose is sufficient for most people, revaccination is recommended at more frequent intervals for people with certain chronic conditions. Check with your doctor to see how often you should have this vaccine.

This injection is an immunization against the *streptococcus pneumoniae*, a bacterium that can cause pneumonia and meningitis. The injection will not, however, protect you against viral pneumonia. As with the flu shot, you might have some tenderness at the injection site, minor achiness, or a low-grade fever.

Ten: Stay positive. How? Try to accept where you are right now in your life. Take each day one at a time and live in the moment. If you are extremely anxious and depressed, seek help from friends and family or from a professional counselor. If you don't have a sense of humor, develop one. Humor helps you look at a lousy situation from a different angle and often makes coping easier.

You don't have to travel this journey alone. There's an army fighting for you. The staff of The LAM Foundation is constantly seeking ways to help all women with LAM as they struggle with this rare disease. Scientists and researchers are striving to unlock the secrets of LAM, and more and more LAM research is being conducted all the time.

# Chapter Four:

## Potential Problems and Solutions



- 4.1 Pneumothorax and its Management
- 4.2 Pleural Effusion and Chylothorax
- 4.3 Angiomyolipomas
- 4.4 Osteoporosis



## 4.1 Pneumothorax and its Management

What is a pneumothorax? In the simplest terms, a pneumothorax, or lung collapse, is a leak of air (pneumo) from the lungs into the chest cavity (thorax). This air leak abruptly changes the pressure between the chest wall and the lungs and causes the lung to deflate and collapse.

For LAM patients, the answer is a little more complex. In LAM, a pneumothorax is usually caused by the rupture of a cyst just below the surface of the lung. The escaping air, now trapped in the chest cavity, puts pressure on the outside of the lung causing the lung to collapse. Basically, whenever one of your cysts on the surface of your lung bursts and lets air out into the chest cavity, your lung may collapse. Often the collapse is only partial, but if your lung partially collapses, you'll still experience some pain and shortness of breath. When a large pneumothorax occurs and collapses a sizable portion, or *all*, of your lung, you'll likely have a good deal of pain and tremendous difficulty breathing. Two thirds of patients with LAM have had a pneumothorax and many women with LAM have had multiple lung collapses (recurrent pneumothoraces).

### Detection

Generally, a chest x-ray will show a pneumothorax. However, if the pneumothorax is very small, it may be difficult to diagnose. If you've gone to the emergency room with pain and difficulty breathing and if your x-ray doesn't show a collapse, you should explain to the medical personnel that you have a lung disease and that pneumothoraces are a common complication of LAM. If necessary, request to see the attending pulmonologist. Ask him or her to read your x-ray and to examine you. Quite often, a small collapse is very difficult to detect on a normal chest x-ray. You may need a CT scan.

### Types and Causes of Pneumothorax

There are two types of pneumothorax: primary and secondary. A primary spontaneous pneumothorax can be caused by a traumatic event such as a penetrating or blunt chest injury. At other times, a pneumothorax can occur spontaneously for no apparent reason.

A secondary spontaneous pneumothorax is one that is usually associated with an underlying lung disease. Diseases affecting the lungs (like LAM and tuberous sclerosis, and other problems such as emphysema and cystic fibrosis) can weaken the structure of the lungs and airways, permitting spontaneous pneumothoraces. An unexplained pneumothorax—or a series of them—often triggers the search for an underlying lung disease like LAM.

### Intervention and Treatment

There are two goals in treating a pneumothorax. The first is the immediate goal of removing the air from the space between the lung and the chest wall (the pleural space) so that the lung can inflate again. The second, and just as important, objective is to prevent recurrent pneumothoraces. These two goals must be weighed when a treatment is chosen.

### Observation

A small pneumothorax (i.e., one involving only 10-15% of the lung) may not require much intervention. Often just bed rest and oxygen are necessary to resolve the problem. Some physicians think that using high concentrations of supplemental oxygen is helpful and sometimes, oxygen may just be used for comfort to make it easier to breathe. Doctors frequently recommend that you allow some time for a small pneumothorax to resolve itself

before you seek more advanced treatment. Remember: a collapsed lung can be dangerous, so don't wait too long to seek treatment, especially if you're really uncomfortable.

## **Chest Tube**

If the pneumothorax is more than minimal in size or doesn't rapidly resolve on its own, the next level of treatment is to insert a catheter (or chest tube) to remove the air. In this process, called a chest-tube thoracostomy, a small hollow plastic tube is inserted into the chest wall, between the ribs, into the pleural space to allow the air to escape. Removing the air, in turn, re-expands the lung. The procedure is generally performed with local anesthesia. Discomfort or pain is common, and risks of the procedure include bleeding and infection. Although a chest tube is uncomfortable, it's a good, immediate first try at fixing a pneumothorax.

Sometimes, if the air leak is really persistent or if air needs to be removed quickly, the chest tube can be connected to a suction device to expedite air removal. If the chest tube will remain in place for a longer period of time, a chest tube with a Heimlich valve is inserted and then the suction box is not needed. This device has a one-way valve that permits air to leave the chest but not to enter it. A really persistent air leak may need even more intervention.\*

## **Pleurodesis**

Once a diagnosis of LAM is made and a pneumothorax occurs, it is recommended that a pleurodesis procedure be done. It has been proven that once you have a pneumothorax, you will be more likely to have subsequent pneumothoraces.

Pleurodesis is a procedure that adheres the outside of the lung to the inside of the chest cavity to prevent the lung from collapsing again. The procedure may be done by a number of different techniques, and many different names are used. Although pleurodesis isn't a foolproof remedy against future pneumothoraces, it generally diminishes the likelihood of additional ones. Also, even if you do have a pneumothorax after you have had pleurodesis, the lung may only partially collapse the next time, and intervention may not be required.

## **Chemical Pleurodesis**

A chemical pleurodesis works by inserting chemicals or other agents into the pleural space to cause adhesion between the chest wall and the lining of the lung to seal the air leak. Chemical pleurodesis can be done either through a chest tube while you're awake in a hospital room, or it can be done under general anesthesia via surgery. Sometimes a combination of chemical and mechanical pleurodesis is used.

*Talc Poudrage:* One of the most common methods of chemical pleurodesis is performed with a chest tube and surgical talc. Often, when a woman already has a chest tube in place, a talc procedure is recommended. Once the air has been expelled, talc is blown through the chest tube into the chest space, a method called poudrage. The talc acts as a sclerosing agent, that is, something that causes the two sides to harden (or stick, in this case) together. Suction is used during this procedure to remove any remaining air in the pleural space. Talc poudrage often produces a burning sensation in the chest as the area heals, but this procedure tends to be very effective, and it's also less invasive than surgery.

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\* Once a lung has had pleurodesis, placement of chest tubes is much more difficult. Any chest tube inserted after a pleurodesis procedure may need to be inserted with the aid of CT scans or x-rays to guide the placement.

*Other Chemicals:* While talc is generally the preferred chemical for pleurodesis, physicians can also inject other chemicals into the pleural space to adhere the lung to the chest wall. Tetracycline and bleomycin seem to be the most commonly used drugs at this time. Talc slurry, a liquid talc solution, can also be used.

### **Mechanical Pleurodesis**

A mechanical pleurodesis (done manually, in an operating room, by a surgeon inserting his or her hands into the pleural space or through video-assisted surgery technology) roughens up the pleura so that when the abrasion heals, the lung adheres to the chest wall. This procedure is often done in combination with some type of chemical pleurodesis as well.

### **Pleurectomy**

If pleurodesis doesn't work or if you have recurrent pneumothoraces, you may be a candidate for a full or partial pleurectomy. This surgery involves stripping off the linings of the lung and the chest wall to cause them to adhere together.

### **Surgery**

To perform the procedures just mentioned, your physician needs access to your lungs. Most doctors use one of the following procedures.

#### *Thoracotomy*

Many pleurodesis procedures require a thoracotomy, a general term for open-chest surgery, a type of surgery that is used for many reasons and procedures. While the patient is under a general anesthesia, the doctor makes an incision that runs approximately from the front to the back (by the shoulder blades) of the chest in between two ribs. This incision allows the doctor access to the lung. Thoracotomies are used in mechanical pleurodesis, in some chemical pleurodesis procedures, and for a pleurectomy.

A patient usually requires about a weeklong stay in the hospital after a thoracotomy but full recovery can take several months. During recovery, the patient should practice deep-breathing techniques to help prevent pneumonia. As with any surgery, bleeding and infection are always risks. Always remember to take whatever pain medications you require.

#### *Thorascopy or VATS*

Video-assisted thorascopic surgery (VATS) is a less invasive procedure than a thoracotomy. VATS is performed by using a fiberoptic scope. Instead of one large incision, you will have several very small ones, which are used to insert the scope and other surgical instruments. The smaller incisions, unlike one large surgical incision, allow a quicker recovery in many cases. For these reasons, VATS has become the preferred method for surgical lung biopsies and other lung surgeries whenever possible.

While pneumothorax and recurrent pneumothoraces are very common in LAM, every woman is affected differently. Talk with your doctor about the options for your individual situation, focusing both on the immediate treatment and the options for how to prevent recurrence.

## Recovery from Treatments

Try to avoid pain after all of these procedures. When your doctors offer you pain medication, take them if you need them. It's much more difficult to stop pain after it has become intense than it is to prevent pain in the first place. You may be in a terrific amount of pain for a few days, but after the first few days, your pain should be manageable. The pain and discomfort should continue to decrease over a few weeks.

You may also experience shortness of breath and exhaustion for a week or two following your release from the hospital, but you may need a month or more of rest before you're able to return to work. Don't push yourself. You don't want to cause another lung collapse by not recuperating sufficiently. Although it may take several weeks to fully recover, you'll probably be able to resume some of your regular activities more quickly. Don't drive until all of your pain is gone and your reaction times are back to normal.

Once the healing is complete, there are usually no residual effects even though your breathing may feel strange at first as if something has changed. Unfortunately, discomfort or a pulling sensation can continue for months after pleurodesis.

### Tips for an Easier Recuperation

- Sleep in a recliner for a few days or for a couple of weeks. The semi-upright position may help because trying to raise the upper part of your body to get out of bed can be painful.
- Have someone buy you a bra that is larger than your usual size. You'll probably need support for a few days after pleurodesis, but your chest may be a bit too swollen from the procedure for your regular bra to fit.
- Don't lift heavy objects until your doctor allows you to do so.

### Transplant after Pleurodesis

Many women have been told that lung transplant after pleurodesis is impossible. While that may have previously been the case, **it's no longer true**. Many women with LAM have undergone successful lung transplants after having every conceivable type of pleurodesis. However, transplant surgery *is* more complicated after you're had one of these procedures.

While pleurodesis no longer rules out transplant, a lung that has been adhered to the chest wall is more difficult to remove, and removing it, therefore, takes more time. The longer a surgery—any surgery—takes, the longer you're under anesthesia and, in this case, the greater your risk of excessive bleeding. However, studies have shown that success rates for women with previous pleurodesis surgery are the same as those without.

Having a pleurodesis can greatly increase your quality of life. By preventing further pneumothorax, you will reduce trips to the ER and stays in the hospital. Talk with your doctor about the risks and benefits to make the best decision for you.

## 4.2 Pleural Effusion and Chylothorax

As you already know, LAM affects each woman quite differently. Some women have no symptoms other than breathlessness, while others struggle with numerous pneumothoraces (lung collapses) or angiomyolipomas (kidney tumors). Many LAM patients are bothered by

pleural effusion, a leakage and accumulation of fluid in the chest cavity. Some have this problem at the time of diagnosis, while others develop it during the course of their disease. Generally, chyle is the fluid that most affects LAM patients, so this chapter will focus on chyle and the problems related to it.

## **Chyle**

Chyle is a milky white fluid consisting of lymph and droplets of triglyceride fat and can sometimes be tinged with blood. Everyone produces chyle (about two or more liters a day); it's a product of digestion. Chyle is normally routed through the lymphatic system and drains into the circulatory system. In LAM patients, the proliferation of smooth muscle cells can damage or obstruct the lymphatic vessels that carry chyle, causing them to weaken and leak. When chyle leaks into the chest cavity, it's called a chylothorax. You can remember it by breaking the word down into parts: *chyle*, "fatty fluid," and *thorax*, "chest"—chyle in the chest.

## **Causes of Chylothorax**

There are many causes of chylothorax, including postsurgical complications and trauma to the body. Sometimes a chylothorax can occur spontaneously, due to the damage that a disease such as LAM can cause to the lymphatic system.

## **Problems with Chyle**

Lymphatic dysfunction in LAM is varied. The most common problem resulting from a chylous leak is chylothorax, a condition resulting from leaked chyle that accumulates around the lungs. The chyle itself is not the problem, but rather the build-up, which causes pressure in the pleural space (around the lungs) and creates breathlessness. Chyle can affect other parts of the body too:

- *Chylous ascites*: chyle in the abdomen
- *Chyloptysis*: coughing up chyle
- *Chyluria*: chyle in the urine
- *Chylous pericardial effusion*: chyle around the heart

## **Symptoms and Detection of Chyle**

If you are having any sort of drainage that is milky or cloudy or if you experience sudden and prolonged shortness of breath, you might have a problem with chylous leakage. You may notice that your belly and/or your extremities swell as the day progresses or you may experience chest pain, a feeling of fullness in the chest, a cough, fever or chills. You may simply feel very, very tired because of the burden the abnormal chyle accumulation is placing on your body. Your doctor may recommend any one of several tests that can help detect abnormal chyle build-up: careful daily measurements (e.g., of your weight or your waistline), a chest X-ray, an abdominal/pelvic ultrasound or a CT scan of the body.

## *Possible Treatments*

If the amount of chyle is small, no treatment may be needed. If the fluid build-up is substantial and causing symptoms, then treatment may be needed.

## *Diet*

You may be told to follow a special low-fat diet. Your doctor, a nutritionist or the experts at NIH can provide one designed just for you. The diet will limit your intake of certain fats, in

particular. Sometimes diet alone is sufficient in controlling chyle, and as an added bonus, it can contribute to your overall health.

If the chyle control is taking too many valuable nutrients from your body, you may need some supplemental oral or intravenous nutrition to give you the nutrients you need. Intravenous treatment gives your gastrointestinal system a break and sometimes provides sufficient time for a damaged, leaking lymph duct to heal.

### *Drainage Treatments*

If the chylous effusion continues in spite of careful diet, you may need to undergo a procedure called a thoracentesis, which is also known as a fluid aspiration from the chest or a pleural tap. In order to drain the accumulated chyle fluid from the pleural space around the lungs, a small needle is temporarily inserted in the chest area from your lower back between your ribs. Sometimes up to a liter of chyle or more can be drained. This procedure is highly effective but may provide only temporary relief. Your doctor will want to assess how fast the chyle builds up again. Some women need to have chyle drained regularly. If chyle production is severe, a chest tube may be inserted into the side of your chest to allow for continuous drainage for several days or longer.

### *Pleurodesis*

If drainage treatments are inadequate, you may be a candidate for either mechanical or chemical pleurodesis, the same procedures used to repair a pneumothorax (see the chapter on Pneumothorax and its Management). For mechanical pleurodesis, the surgeon abrades the surface of the lung and the inside lining of the ribcage to cause adhesions between these two surfaces. During chemical pleurodesis, a chemical agent such as talc is introduced into the pleural space either through a chest tube or during surgery.

### *Surgery*

Finally, if you need further help controlling chyle, you may have surgery to tie off a structure called the thoracic duct. This procedure is called a thoracic duct ligation. This duct can be difficult to locate, and your surgeon may employ VATS (use of a scope and video camera to look into the chest) or thoracotomy (open-chest surgery through a large incision on the side of your chest). Other less commonly used surgical options for management of chylothorax include the removal of the lining of the lung (pleurectomy), or, rarely, placement of a pleuroperitoneal shunt (a tube placed inside the body that connects the chest cavity to the abdomen to drain fluid from the chest).

### *Medications*

Rapamune has proven to be effective in the management of chyle for some patients. Check out section 1.3 to learn more about treatment with Rapamune.

## **4.3 Angiomyolipomas**

Angiomyolipomas are benign tumors that are found in the kidneys of about 40% of women with LAM and in the majority of women with tuberous sclerosis. Although extrarenal (outside the kidneys) Angiomyolipomas are rare and are sometimes found in the liver, uterus, lymph nodes, blood vessels and in other sites throughout the body.

As with many medical terms, the word *angiomyolipoma* describes the tumor itself. Angiomyolipomas are composed of three types of tissue: *angio* “blood vessels,” *myo* “smooth-muscle cells,” and *lipo* “fat or adipose tissue.” Add to these terms the suffix *oma*, meaning “tumor,” and you have an *angiomyolipoma*.

The amount of each type of tissue can vary greatly with each tumor, and this variance can determine if the angiomyolipoma will produce any symptoms. For example, a tumor with more blood vessels would probably be more prone to bleed and could possibly grow faster than one with a higher percentage of fat.

Angiomyolipomas can usually be detected without an invasive procedure. Fat (often our enemy but in this case our friend), due to its density, shows up black on an x-ray, and white on an ultrasound. Fat is the component that generally allows these tumors to be diagnosed with a high-resolution CT scan or ultrasound. Your radiologist cannot be 100% certain that your tumor is an angiomyolipoma though, because some cancerous tumors also contain a large quantity of fat. If he or she is aware that your diagnosis is LAM, the odds are high that your tumor is a benign angiomyolipoma. As you read on, remember that only 40% of women with LAM have angiomyolipomas and that many of those tumors never cause any problems.

### **Guidelines for Treatment**

You’re probably wondering what to do about these tumors. Some general guidelines are given below and these include waiting and watching\*, embolization or surgical removal of the tumor. This information is provided to give you some background as to what to expect when you discuss your situation with your doctor. But keep in mind, these are just general guidelines. As with most health concerns, be sure to follow your doctor’s recommendations for you.

- If a tumor is less than 4 centimeters, or about 1½ inches, (the size refers to the largest dimension of the tumor) and if it is not causing you any problems, you can usually monitor it with annual CT scans or renal ultrasounds.
- If the tumor is larger than 4 or 5 centimeters, you might want to have it checked every six months.
- If the tumor is less than 4 centimeters and is causing you pain (usually in the lower back), if it’s hemorrhaging (you might notice blood in your urine) or if you are nauseous, consult your doctor immediately. After the two of you assess your situation, you might feel comfortable waiting a few days to see if the problem subsides.
- If the tumor is over 4 centimeters and is causing you any of the above problems, you should seriously consider having it embolized or surgically removed.

For LAM patients, some doctors are now recommending embolization or the removal of any tumors over 5 centimeters. Take this recommendation seriously. The larger a tumor grows, the greater the chance that it’ll cause you problems by bleeding. Of patients who have such bleeds, a quarter of them bleed significantly and such bleeding can be life-threatening. If you have these tumors removed when you are healthy, you will recuperate much more quickly. Should you wait, for example, to have a large tumor removed until you’re listed for transplant, you might be removed from the active transplant list while you recover from the surgery or

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\* The growth rate of these tumors is quite variable and unpredictable, so your tumor may grow extremely slowly or very quickly. Just make sure to have it monitored regularly.

embolization. Think twice, and then a third time, before you put off a procedure that your doctor recommends.

## **Embolization**

Embolization is a process that cuts off the blood supply to the tumor. The procedure can be performed to prevent a rupture or hemorrhage of a large AML, or it can be used to stop an AML that is already bleeding.

An embolization is much like a heart catheterization. Doctors, usually angiographers (people trained to go into the blood vessels with help from an x-ray), go into an artery in your groin. They thread a catheter up to your kidney and then into the blood vessel supplying the tumor. By injecting a contrast dye into your veins, they can see which blood vessel feeds the tumor. The blood supply is cut off by injecting tiny polyvinyl particles into the artery (like building a dam) or by another means of constricting the vessel.

The procedure is relatively painless, but you will need to lie flat for about six hours afterward to give your artery a chance to close up. You may be given antibiotics or a steroid through your IV to help reduce infection, pain and/or fever that may result as your body responds to the embolization procedure.

Following the procedure, if you have much flank pain or a rather high fever, you may be suffering from Postembolization Syndrome (PES). This term refers to a severe response of the body to the embolization procedure, possibly due to the body's immune response to the dead tissue from the blocked tumor. About 85% of people who have angiomyolipoma embolization will experience this but a low dose of prednisone or other steroid can alleviate the problem.

Once a tumor is embolized, it takes several months or more for the angiomyolipoma to shrink up, die and be absorbed into the body. Unfortunately, the tumor may recur if its blood supply is not completely closed (or if the tumor is not totally removed during surgery).

Embolization is advantageous in several ways. First, the procedure is much less invasive than surgery. Second, because it's less invasive, most patients tend to recover quickly. Third, because the procedure is often done as an outpatient surgery or, at most an overnight stay, the cost is generally less. Shorter hospital stays are cheaper than longer ones. Shorter recuperation time means you—or your caregiver—can get back to work more quickly.

Most often embolization can be done but on rare occasions, surgery is necessary. The size of the tumor, its position and its involvement with the kidney are all factors that determine whether embolization or kidney surgery is right for you.

## **Surgery**

If you and your doctor decide that you require surgery, talk to the surgeon beforehand. You may need to have the entire kidney removed in a procedure called a *total nephrectomy*, or you may have just a part of the kidney removed, a *partial nephrectomy*. Let your surgeon know that you would like to have as little of your kidney removed as possible. Because the treatment for end-stage LAM is lung transplantation and because kidneys are essential in processing powerful immunosuppressants after transplant, you need as much of your kidney as your surgeon can save. But you must also understand that you might need to have the entire kidney removed. If this is the case, don't lose heart. Many people live long and productive lives with only one

kidney. And nowadays, having only one kidney does not automatically disqualify you for a future lung transplant. Work with your surgeon to decide if a total nephrectomy is necessary.

Angiomyolipomas can be a pain, both literally and figuratively. They can make your life more complicated. But keep in mind that these tumors are one aspect of LAM that can be treated and can be improved. It is important to understand your options and work with your doctor to minimize the problem as best you can.

#### **4.4 Osteoporosis**

Many women who have LAM also have osteoporosis. What is it? It's a disease that decreases your bone mass. The bones either stop building bone mass or they lose what mass they have and become less dense and more brittle. In simple terms, that means that your bones become more fragile and more likely to break. Another condition known as osteopenia, the loss of bone mass below normal levels, is related to osteoporosis. With osteopenia, the bones are beginning to lose mass. As osteopenia worsens, it becomes osteoporosis. Because osteoporosis is usually an invisible disease, you probably won't discover it until you break a bone. Ask your doctor for a bone-density scan\* to see if your bones are losing mass.

Osteoporosis is a well-known problem, especially among older women, throughout the world. As women age, they lose bone mass because they have less estrogen in their bodies. Estrogen helps keep bones healthy in women. However, even young women with LAM are at risk to develop osteoporosis. Why? No one has the full answer to that question yet, but there are some contributing factors. First, many women with LAM have used hormonal treatments like progesterone or have had their ovaries removed, which induces menopause, to slow their disease. These treatments, due to their anti-estrogen tendencies, cause bones to lose density. Second, anti-inflammatory drugs like Prednisone and other steroids, which many LAM patients use to help them breathe more easily, tend to decrease bone mass by interfering with calcium absorption. Third, because women with LAM often have trouble breathing, they often neglect exercise, especially weight-bearing exercise, which helps to keep bones healthy. Add these to other risk factors for osteoporosis—smoking, heavy alcohol use, a family history of osteoporosis and a diet low in calcium—and it's easy to see why many women struggle to maintain bone density.

##### **What Can You Do about It?**

One of the first things you should do after being diagnosed with LAM is to get a bone-density scan. Your first test will act as a baseline to let your doctor know the initial condition of your bones. You may already have osteopenia or osteoporosis, but by having the scan, you might be able to keep the problem from worsening.

If the scan shows your bones are fine, you should monitor them with regular scans about a year apart or as often as your doctor recommends. If you're in the protocol at NIH, you'll receive a bone scan yearly.

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\* May also be called a bone-mineral density test.

## Medications

If you do have either osteopenia or osteoporosis, your doctors will probably put you on a regimen to increase your bone density. They may recommend prescription medicines to help prevent bone loss and to rebuild the bone structure. Actonel (risedronate) or Fosamax (alendronate)\* are the two most common oral (taken by mouth) drugs prescribed and both come in daily or weekly doses. Many women prefer the weekly dose because taking either drug can be a bit of a bother. You must take the drug on an empty stomach with a full glass (about eight ounces) of water when you first wake up. You cannot take any food or any other beverages—not even your first sip of coffee—for thirty minutes. You also need to remain upright—sitting or standing—for that length of time. No going back to bed for a few more minutes! Following these guidelines is very important; if you don't drink enough water, if you eat or drink anything else too soon, or if you lie down, the drug can irritate or even burn your esophagus. Some doctors even recommend that you stay upright and refrain from eating and drinking a bit longer. These strict rules convince many women to take the drugs just one day a week. Others find that the weekly dose is a bit too strong for them, and they prefer to take the drugs daily. For them, the routine becomes as common as brushing their teeth.

If Actonel and Fosamax don't seem to work for you, try Miacalcin (calcitonin-salmon), a nasal spray prescribed for osteoporosis. Some women prefer it because it enters the bloodstream rather than the digestive track and, therefore, doesn't cause esophageal or stomach problems.

Your doctor may also recommend additional calcium in the form of supplements. Depending on your diet, you may take 1,000-1,300 milligrams of calcium daily, depending on your age and whether or not you're pregnant, nursing, or in menopause. Many calcium tablets come with added vitamin D to help the body absorb the calcium. If your calcium supplement doesn't contain vitamin D, be sure you are taking a multivitamin supplement as well. Many doctors recommend 400-800 IUs (international units) daily of vitamin D. Follow your doctor's orders as to when, how much, and how often to take the supplements.

## Diet

If you're taking medications or even if you don't need them, you can always beef up (maybe "milk up" would be a better term) your diet with calcium. An extra glass or two of milk, some hard cheese, a cup of yogurt or cottage cheese, or a bowl of ice cream can add much needed calcium to your diet. Just choose the lower-fat versions so you don't increase your weight while you're increasing your bone mass. Dark green leafy vegetables like turnip greens, collard greens, kale, and broccoli are also good sources of calcium as are shrimp, salmon, oysters, and clams. And don't just try to build bone mass. Try to avoid losing it too. Smoking and excessive alcohol consumption can strip your body of much needed calcium. So eat smart and drink sensibly.

## Exercise

Exercise, especially weight-bearing exercise, is essential in keeping bones strong. Walking is the simplest and most convenient form of weight-bearing exercise. It's considered to be weight bearing because your feet are supporting the weight of your entire body. So, if you aren't already walking about thirty minutes a day, get out and walk. Try lifting lightweights, building

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\* These two drugs are in a class of drugs known as "bisphosphonates."

up to heavier ones. Ask your doctor to recommend some exercises to help get you started, join a gym, or hire a personal trainer. But get moving.

### **Fall-proofing Your House**

If you already have osteoporosis, you'll want to do everything in your power to prevent fractures. Even if you take medications, calcium and vitamin D supplements, eat a calcium-rich diet, and exercise, you should still take other precautions. "Fallproof" your house. Just as you childproofed your house when you had babies, go through your house and see what you can change to avoid falls that might fracture your bones.

Remove area rugs that tend to slide along the floor and cause you to slip. If you need the rug, add a pad underneath that will hold the rug securely in place. Have good lighting throughout the house so that you can always see where you are going. Install nightlights in hallways and in bathrooms for nighttime visits to the bathroom or kitchen. There are inexpensive nightlights that plug into outlets and that come on automatically when it's dark so you don't have to worry about turning them on and off.

Wear clothing that isn't too long. You don't want to trip on your hem and fall down the stairs. Wear slippers with skid proof soles. Don't wear socks without shoes or slippers; socks alone can really be slippery, especially on wooden floors.

Osteoporosis is potentially very dangerous. However, it's one of the problems we encounter with LAM that can be controlled. Be sure that you tell your doctor about the extra likelihood that you, as a LAM patient, could have osteoporosis. It's treatable! Don't ignore it!



# Chapter Five:

## Supplemental Oxygen



- 5.1 Supplemental Oxygen
- 5.2 Oxygen Saturations
- 5.3 Tips for Those Who Need  
Supplemental Oxygen
- 5.4 Traveling with Oxygen



## 5.1 Supplemental Oxygen

You've just found out that you need to start using oxygen. You have visions of dragging around a large steel tank, barely being able to maneuver it. Sound like your worst nightmare? Sure it does. But using supplemental oxygen isn't really a nightmare; oxygen can be a huge enhancement to your quality of life.

The first and most important fact you need to know about supplemental oxygen is that using it will make you feel so much better! If you're often out of breath for a long period of time, if you haven't been sleeping well or if you have no energy—oxygen will probably make a huge difference in your life.

A brief explanation will help you understand why oxygen will help you. Oxygen (O<sub>2</sub>) is an element (remember chemistry class?), a gas your body needs to live and to perform all of the bodily functions no matter how small. In general, when you inhale, you take in oxygen, which then passes through your lungs and into your blood through alveoli, small air sacs in the lungs. Once your blood has picked up the oxygen, it's pumped out to the rest of your body. As your body uses the oxygen for its various functions, it releases carbon dioxide (CO<sub>2</sub>) as a waste product. The carbon dioxide, in turn, travels through your blood into the cells in your lungs, up through your lungs, and you breathe it out. If this process is flawed and your body can't pass oxygen from your lungs into your blood, the oxygen levels in your blood will not be sufficient for your body to function well and you may need supplemental oxygen. This is especially true for people who are unable to obtain sufficient oxygen due to respiratory illnesses like LAM.

### Why You Should Wear Oxygen

Okay, let's face it. You don't really want to wear oxygen. You get out of breath, but you recover quickly, so you don't think you're ready for supplemental oxygen. However, tests which are less subjective than your feelings may show that you're damaging your body by not using O<sub>2</sub>. Lack of oxygen (hypoxia) can make you feel tired, weak, confused and/or forgetful. But a more serious problem is that chronic oxygen deficiency can force your heart to work harder than is healthy for it. This constant strain on your heart can cause pulmonary hypertension (when the blood pressure in the pulmonary artery is abnormally high) and other heart problems. These problems are exacerbated because, even though you usually know when your lungs don't get enough oxygen, you can't tell when your heart and your brain aren't getting enough oxygen. Are you willing to risk a heart attack because you don't want to use supplemental oxygen?

You can take a few simple tests at your pulmonary center or at the National Institutes of Health (NIH) to determine your actual oxygen needs. If you're fine without oxygen, that's good news. But you may not realize how run-down and exhausted you've become simply because you've gotten used to feeling that way. Once you start using supplemental oxygen, you'll regain much of your lost energy and you'll be able to be more active. Furthermore, you'll have the security of knowing that you're not straining your heart.

There are a few myths circulating about oxygen usage. One of the most common ones is that you can become "dependent" on oxygen or "addicted" to it. This just isn't true. Another myth is that you can store enough oxygen when you're wearing it while resting at home, for example, to "tide you over" during more active periods. The reverse doesn't work either—you can't use it as a "rescue therapy" to give your body more oxygen after a period when you know your body wasn't getting enough. If your body needs oxygen, it needs oxygen right then. Not

before to prepare for a deficit and not after to make up for one. By not using oxygen when you need it, you're only cheating yourself. One more myth is that oxygen tanks can "blow up" or explode. The tanks are under pressure, but if the top is broken off a tank, the tank won't explode but it will soar across the room like a rocket (don't try this at home) until it's either empty or hits something.

There is one thing that's *not* a myth: There should be **NO SMOKING** around oxygen. The tank won't explode but oxygen fuels fires and if you have supplemental oxygen flowing around your face, everything around you will be oxygen enriched. Therefore, even a spark will have plenty of oxygen to burn more quickly into a larger fire.

When you begin using O<sub>2</sub>, your doctor may prescribe it for you only to use when you're sleeping or exercising. In general, your lungs—like other parts of your body—slow down during sleep. Many women and men (especially those who are overweight) tend to snore, suffer from sleep apnea or have other conditions that cause their oxygen levels to fall during sleep. Therefore, you might not get the amount of oxygen you would need even if you didn't have LAM. Your doctor can order a test called an overnight oximetry, which can be done at home or in a sleep clinic and which measures your oxygen levels during sleep, to determine if you need nighttime oxygen. Sometimes, O<sub>2</sub> at night is all you need to keep your oxygen levels up to a sufficient level.\*

O<sub>2</sub> usage may vary according to your individual needs and your level of activity. While sitting to read or watch TV, you may require less oxygen than when walking or vacuuming. Some women desaturate (that is, the oxygen level in the blood drops) significantly when they are exercising. For them, O<sub>2</sub> during workouts, heavy housework, or gardening may fulfill their oxygen needs. Your body doesn't "store" oxygen for the times when you really need it, so when you need it, use it.

It's important to remember that oxygen needs can, and do, change as LAM progresses, so you'll need regular checkups to monitor your oxygen levels. How will you know if you need oxygen? Your doctor will tell you based on your O<sub>2</sub> saturation levels and on your PFTs. But in general, if your oxygen levels are below 90% at rest, you'll probably need full-time oxygen. If your saturation levels drop below 90% only during sleep or exertion, you'll only need oxygen at those times,\* but many women find that they eventually require oxygen full-time.

Before you begin to use supplemental oxygen, you'll need your doctor or your respiratory therapist to determine how much oxygen you require. Since oxygen flow is measured in liters per minute (LPM), you need to know how many liters of oxygen you require per minute to keep your O<sub>2</sub> saturation levels up to a certain percentage (usually 90% or higher). Your liter flow will differ with different activities.

Acquisition of supplemental oxygen requires a prescription from your doctor. Health insurance often covers a significant percentage—and sometimes all—of the cost.

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\* Most doctors say you should keep your oxygen level at 90% or above.

\* Some doctors refer to an S curve, for a visual representation, when they described how your saturation levels drop. Picture the letter S. Draw the S from the top right to the bottom left. As you draw the first or top segment, think of that part as equal to 90-100% of your oxygen saturation. The entire segment is a relatively straight line. So if you drop from 100 to 92%, for example, the change is slight. But once you drop below 90%, the S begins to curve downward quickly, and this sharp decline shows how quickly your oxygen saturation begins to drop. And once you get below 90%, your heart begins to work much harder.

## Oxygen Delivery Systems

### *Compressed Gaseous Oxygen*

You've seen people lugging around those big steel or aluminum tanks of oxygen. These tanks carry compressed gaseous oxygen, a relatively inexpensive form of oxygen. Although these tanks are rapidly becoming outdated, they may be all your insurance company will provide, especially if you only use tanks occasionally. The tanks come in a variety of sizes so you can order different sizes of tanks and use them to suit your needs.

The tanks vary in size from small to large. The smallest tank, a "B" or "M6" size, weighs about three pounds and lasts about an hour on two liters of oxygen per minute without an oxygen-conserving device (discussed a little later). The next larger size, "C" tanks, weighs about six pounds and lasts over an hour on four LPM. "D" tanks weigh about eight to ten pounds (still small enough to carry in a backpack), are taller than both "B" and "C" tanks, and last about an hour or two, depending on your use. "E" tanks are quite heavy—about 12 to 15 pounds—and need to be pulled in a trolley or cart. These tanks last about two hours and fifteen minutes (without a conserving device) if you're using four LPM of oxygen.

Keep in mind that all of these times are approximate. Individual usage varies, and you need to learn (probably by trial and error) how long various size tanks last for your usage. Despite the versatility of tank sizes, these gaseous oxygen tanks are inconvenient because you need to keep them on hand and store them carefully. If you travel with tanks, even for short distances, wedge them securely in the back seat of your car. Don't store them in the trunk because of the risk of a rear-end collision. If a tank rolls and the top breaks off, the forceful release of oxygen would send the tank shooting off like a torpedo or missile.

### *Liquid Oxygen*

Liquid oxygen is the most expensive oxygen-delivery system. Liquid oxygen is produced through a process that cools the gaseous oxygen until it becomes a liquid. Many people prefer liquid oxygen because the containers are smaller, more convenient to use, and can last up to eight hours, depending on your usage. Unfortunately, it does have some drawbacks: liquid oxygen evaporates rather quickly, and, unless you're using it constantly, some insurance companies do not pay for this form of oxygen. Also, because it evaporates quickly, you shouldn't store your supply near a heat source.

Liquid oxygen is generally delivered to your house in a large tank, called a reservoir, which is refilled on a regular basis—weekly, biweekly, or monthly, depending on your use. Your oxygen company also provides smaller portable canisters to fill from this large one.

### *Concentrators*

Concentrators are large (most weigh about 50 pounds), and sometimes noisy, machines that extract oxygen from room air, eliminate other gases, and concentrate the oxygen, sending it into your tubing. Concentrators provide you with an inexhaustible supply of oxygen (as long as the electricity is working). Although not easily portable, concentrators are a great option for anyone using oxygen at home. If you have an oxygen concentrator, you'll also be supplied with compressed gas or liquid oxygen for a backup should the electrical power fail in your home and for your portability needs.

A concentrator, positioned in a central location and fitted with long tubing (usually fifty feet will get to all the corners of your house unless you're living in a mansion), allows you to reach almost any part of your house without carrying around an oxygen tank.

Unlike gaseous tanks and liquid-oxygen canisters, your concentrator is a machine that needs some care to keep it from breaking down. Keep it away from heating vents so it doesn't have to work too hard and clean the filters regularly so that dirt doesn't obstruct the airflow. And for best results, put the concentrator in a large room so it has more air to draw from.

If you believe that your concentrator is not working properly even if the alarm on it does not go off, call your oxygen supplier and ask the company to send someone out to check the machine. Many suppliers replace concentrators yearly, taking the old ones back to the company for routine maintenance. But even if you've just received another concentrator, there is no guarantee it will continue to work correctly.

Some concentrators, especially older models, are quite loud. If the noise bothers you, place the concentrator in another room or set it on carpeting or even on a large piece of thick cork; both materials work as noise reducers.

Choosing the "best" oxygen-delivery system for you will depend on how much oxygen you require. For low flow requirements (2-3 LPM), a concentrator at home and compressed gas cylinders for travel may be the most economical. For high flow requirements (over 5 to 6 LPM), a home concentrator may not deliver a sufficiently high liter flow, and a liquid oxygen system may be required. If you spend a lot of time away from home, a liquid-oxygen system may provide you with the smallest amount of equipment for the largest amount of time, even if your oxygen-flow requirements are fairly modest.

## **Getting the Oxygen into Your Lungs**

### *Cannula*

A cannula (pronounced can-yoo-luh) is that funny-looking circular piece of tubing with prongs that fit into your nostrils. The cannula attaches to a longer piece of tubing that is hooked up to your oxygen supply. Generally, you put the prongs into your nostrils, take the tubing up and over your ears, and tighten up the slack a bit under your chin (like the string on the cowboy hats you wore when you were a kid). Some women, however, swear that it feels better to put the tubing over their heads after inserting it into the nostrils and to adjust the slack at the base of the hairline in the back.

Cannulas come in different materials, sizes, and shapes. Some are made from hardened plastic while others are made from silicon. Some cannulas are latex-free due to a high number of people with allergies to products made from rubber. Even the length and diameter of cannulas can vary. For instance, you can get four-foot or seven-foot cannulas to wear with your portable tanks, while your cannulas worn at home may be only a foot long since you attach them to fifty feet of tubing. In the same manner, the diameter of some cannulas may only accommodate a liter flow of three liters, and others will support six liters or more.

The nasal prongs on some cannulas are straight while others are curved. Some prongs are spaced farther apart than others. Even the tips of the prongs vary. They can be straight, tapered or flared. If the cannula you normally wear is uncomfortable or is bothering you for

some reason, talk to your respiratory therapist or your oxygen supplier. The solution may be as simple as cutting off a sliver of the prong tips to make them fit in your nose better. Or you might have to experiment with different styles of cannulas to find the one that fits you best. But there is no reason for you to be uncomfortable.

Use a new cannula each week. Pick a day of the week, and change your cannula regularly on that day so you will always remember. If the smell of new plastic bothers you, remove your cannula and tubing from their packaging twenty-four hours prior to use to “air” them out. The smell will lessen considerably or disappear completely.

### *Mask*

A lightweight mask is recommended for high flow needs (usually over 6 LPM). The mask fits snugly but comfortably over your mouth and nostrils and maximizes the amount of O<sub>2</sub> that can actually enter into your lungs. When used along with a nasal cannula, the efficacy of O<sub>2</sub> delivery is even greater.

It’s important to change your mask weekly as you would your cannula.

### *Transtracheal Oxygen Delivery*

If you aren’t receiving sufficient oxygen through a cannula or a mask, transtracheal oxygen delivery is another alternative. This delivery system is very efficient because all of the oxygen enters your lungs rather than having some escape from your nose or mouth. A surgeon makes a small hole in your trachea (just as he or she would for a tracheotomy, but the hole for a tracheotomy would be much larger). Once the procedure is done, the patient inserts a catheter into this hole and attaches the catheter directly to the oxygen supply. Usually this method can reduce your liter-flow requirements. Another benefit of this method is that you will not experience nasal irritation or dryness because you’re not using a nasal cannula.

Just as you would change your cannula on a regular basis, you also need to care for this tracheal opening, which needs to be cleaned twice a day. You remove the catheter (a process called stripping), clean the site (special solutions are available), and place a clean catheter (one you’ve soaked in a sterile solution) in the opening. The process may sound really gross, but some women find a transtracheal catheter the most efficient and least distracting delivery system. Don’t be hesitant about using it because you are afraid of the responsibility of removing the catheter and cleaning it. Transtracheal oxygen delivery may be the method you *need* to receive sufficient oxygen.

If you choose this system, you can wear turtleneck sweaters, shirts with higher collars, or pretty scarves to hide the tracheal opening. You can also hide the tubing inside your clothes, letting it hang out at waist level. You’ll still need to connect your tubing to a concentrator or a tank, but some women feel less conspicuous with a transtracheal supply system than they do wearing a cannula.

Finally, when you no longer need oxygen (e.g., after transplant), the hole usually closes up on its own. If it doesn’t, only a couple of stitches are required to close it.

To learn more about transtracheal oxygen delivery, check out the Transtracheal Systems website: <http://www.tto2.com>.

## *Tubing*

Tubing, made of latex-free plastic, connects your cannula to whichever delivery system you choose to use—compressed gas, liquid oxygen or a concentrator. Generally, cannulas used with compressed air and liquid oxygen are pre-attached to tubing that is either four feet or seven feet long. A four-foot cannula is best if you'll be carrying your own oxygen supply because you're less likely to trip on it. A seven-foot one is better if you're wheeling your oxygen in a cart or if someone else is being kind and carrying your oxygen tank for you.

When using a concentrator, 25 feet and 50 feet of tubing are the standard lengths. Fifty feet of tubing can reach most places in most houses if the concentrator is placed in a central location. But if you have a small efficiency apartment, twenty-five feet of tubing might be perfect—enough to move around easily without tripping over excess footage.

Most tubing is clear but green tubing is available. The green plastic stands out visually. If you have problems constantly tripping over your tubing, request green tubing and see if it helps reduce the number of times you trip.

## *Regulators*

Regulators are devices that attach to the tops of compressed-gas oxygen tanks. They have gauges that indicate how much oxygen is in your tank. They also have a liter-flow valve (basically an on-off switch with a volume-like control) that dispenses the oxygen flow you require. The regulator fits up over the top of your tank and you tighten it onto the tank. Once the regulator is attached, you open the tank with either a key-like device or a knob, if the tank is larger. Once the valve is open, you can turn on the flow of oxygen and adjust it to suit your needs.

Regulators are very easy to use, but if you hear a hissing sound even before you open the liter-flow valve, air is leaking out of your tank. Try attaching the regulator again and if you still hear the hissing, check the O-ring on your regulator. This is a small rubber, plastic or rubber and metal ring that is used as a gasket and that is usually attached to the regulator. The O-ring is necessary to make a tight seal between the regulator and the tank. If you have a leak, turn off the regulator, shut the valve on the tank and remove the O-ring. Flip the O-ring over and try again. If your tank continues to leak, you may need a new O-ring or you may need a new regulator. Many compressed air tanks now have built-in O-rings, a feature that eliminates this problem.

## *Oxygen-Conserving Devices*

When oxygen is delivered in a continuous flow, much of the oxygen is wasted because you're exhaling part of the time. Oxygen-conserving devices try to provide oxygen in more efficient ways, allowing you to get more time from your tanks. These devices are divided into two main types: conserving cannulas (e.g., Oxymizers) and conserving regulators (e.g., Pulse Dose)\* regulators. A conserving cannula is a larger cannula with either a pendant or "mustache" reservoir or chamber attached to it. With this type of device, the oxygen is held temporarily in the reservoir until you take a breath.

If you're on a high liter flow rate of oxygen, a conserving cannula can cut the flow rate almost in half. For example, if you need 5 LPM per nasal cannula, you may be able to use 3 LPM

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\* Both Oxymizer and Pulse-dose are brand names.

per oxymizer. The puff of air coming from such a device will usually create a small but audible sound.

Another type of conserving device, called a conserving regulator, attaches directly to your oxygen tank in place of the normal regulator. With a regular system, oxygen is continually flowing into your nostrils. When you use this type of device, oxygen is only released to the tubing when you inhale. Inhalation will bring the air you need, along with a slight click and a whooshing sound. Conserving regulators can almost double the time a tank will last.

### *Nasal Dryness*

Nasal dryness seems to bother most people who wear oxygen, although those who use liquid oxygen tend to have fewer problems with dryness. Humidifiers can be attached to your concentrator (or other oxygen supply) to help reduce such problems. Your oxygen company will show you how to hook one of these to your machine. If dryness persists, try using a saline solution to irrigate the nasal passages. These solutions can be purchased without a prescription at any drug store and at other stores too, often in the allergy-remedy aisle.

You can also rinse your nose with a neti pot, a ceramic pot that looks like Aladdin's lamp. Mix one teaspoon of kosher salt with warm water in the pot and hold your head over the sink. As you pour the salt water into one nostril, it flows out the other. Repeat the process, pouring into the other side. (It's easier than it sounds.) The salt water rinses your nose and soothes and moisturizes the mucous membranes. Neti pots are about \$15.00 and are available from at the drug store and at many health-food stores.

NEVER use petroleum-based lubricants (e.g., Vaseline) in, under, or around your nose. In the presence of oxygen, petroleum burns. Use water-based lubricants (e.g., KY Jelly) if needed for relief of dryness or chafing from the cannula.

### *Oxygen Delivery through Eyeglasses*

One other method of oxygen delivery is available: through special eyeglass frames. This method is a bit costly, but it's one more option. Using these glasses minimizes the visibility of your cannula.

Special tubing attaches to the earpiece of these unique hollow-frame glasses. The oxygen flows through the frames via the temple pieces, over the top of the frame, and finally through small cannula hooks that protrude down along the sides of your nose and into your nostrils. These glasses have been available for at least five years, but now the selection of frames is much greater.

To learn about this product, check out Oxy-View at <http://www.oxyview.com>.

### **Oxygen Companies**

When the time comes for you to use supplemental oxygen, you should shop around to find an oxygen supplier that fits your needs. This may be easier said than done, however, since many women are restricted to certain companies by their insurance company who will need

documentation to approve this expense\*. But even if you find yourself restricted by your insurance company, you still have some room for negotiation.

You'll most likely want to remain as independent and as active as you can. To do this, you might need several types of oxygen: a concentrator to use when you're at home, compressed air tanks when you're on a long trip and small liquid-oxygen canisters for working or running errands. Or you might even request your supplier to deliver oxygen or a concentrator to both your home and your office so you don't become short of breath hauling oxygen tanks back and forth. These companies will work with you and the equipment is there for you. Find a solution that works best for you.

Finally, if you have a choice in what company you select and if you know you will be traveling, choosing an oxygen supplier that is a national chain or that has connections across the United States will facilitate your traveling with oxygen. Larger companies such as Apria and Lincare can generally provide oxygen to you more easily when you travel.

### Your Rights – Supplemental Oxygen Guide

Oxygen therapy can be complicated but it's important to know that as a consumer of the oxygen supplier, you have rights regarding the services that are provided to you. It's important to remember that the oxygen supplier is not your doctor and cannot make decisions about the type of equipment you require! The LAM Foundation and the COPD Foundation have worked to put together our Supplemental Oxygen Guide which details what your rights are as a consumer of your oxygen supplier. The guide can be found on The LAM Foundation's website at <https://www.thelamfoundation.org/LAM-Patients-Family-Friends/Resources/Supplemental-Oxygen-Guide>. If you have any questions about the service (or lack of service) that you are receiving from your oxygen supplier, you should contact the COPD Information Line at (866) 316-2673.

### **Getting Used to Oxygen**

Wearing supplemental oxygen in public can be very difficult at first. You need to remember that it will make you breathe easier and that it's only a physical manifestation of your otherwise invisible disease. Do you wear glasses? If you do, you probably hated them the first few times you wore them. But you got used to them, and you'll get used to oxygen too, even though you may always consider it a bit of a burden to cart around. The people you know will become so accustomed to your O<sub>2</sub> that they'll stop noticing it. Other people may stare, but who cares? Haven't you always imagined people were looking at you? *Did I remember to zip my jeans? Do I have spinach in my teeth? Is my blouse gapping?* We all have those insecure worries sometimes. Wear your oxygen proudly. Most people have something they don't like about themselves. Enjoy the freedom oxygen gives you. Look as if you are comfortable with it, and you'll put other people at ease.

Hold your head up high with that hose in your nose. You are doing people a favor by showing them that a physical disability doesn't make you any less of a person. If your sister needed to wear O<sub>2</sub>, would you be embarrassed to be seen with her? Of course not!

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\* Although your doctor will probably want you to use oxygen when your saturations dip below 90%, many insurance companies will not pay for oxygen until you desaturate below 88%. Be prepared to fight for oxygen to keep your body healthy.

One more thing to keep in mind: some people will ask you why someone as young and healthy-looking as you needs O<sub>2</sub>. If you want to shut them up, just say, “Because it makes me look pretty, don’t you think?” But you can also use the question to educate those people about LAM. Keep your informational brochures handy, and be ready to spread awareness!

## 5.2 Oxygen Saturations

Now that you have been diagnosed with LAM, you’ll need to make sure that your body is getting enough oxygen because oxygen saturation levels can fall as your disease progresses. There are several tests that measure your levels.

### Arterial Blood Gas

An arterial blood gas (ABG) measures the percentage of oxygen and carbon dioxide in your blood. It can determine if there is an imbalance (and if there is, how severe it is) between the O<sub>2</sub> and CO<sub>2</sub> levels. LAM can easily cause this imbalance and you’ll need regular ABGs to monitor your condition. The results of the test can tell you if you need supplemental oxygen or how successful your oxygen therapy is.

The blood is taken by drawing a blood sample from an artery (usually the radial one near your wrist or from the brachial artery in the crook of your arm) rather than a vein. This blood draw causes more discomfort than one taken from a vein, and, because the arteries are harder to find, the draw takes a bit more time too.

After the blood has been drawn, the technician will slip the vial of blood into some ice and he or she will then apply pressure to the site where the blood was drawn. It’s very important that the artery seals up to prevent bleeding, so be patient.

The values from this test are the PaO<sub>2</sub> (or PO<sub>2</sub>), the arterial partial pressure of oxygen and the PaCO<sub>2</sub> (PCO<sub>2</sub>), the arterial partial pressure of carbon dioxide. It’s really important that your blood has normal levels of both oxygen and carbon dioxide because if it doesn’t, the rest of your body will need to compensate in some way.

You might hear your doctor mention the pH level of your blood. If you remember your chemistry class, the pH level refers to the degree of acidity or alkalinity.\* Some respiratory problems like LAM tend to make your blood too alkaline if you can’t get enough oxygen in or enough carbon dioxide out or too acidic if you take in too much oxygen and put too much carbon dioxide out. Even if you think the test is a bit painful, it can give your doctor vital information.

### Pulse Oximetry or SaO<sub>2</sub>

This test is a noninvasive procedure to measure your oxygen saturation, that is, the amount of oxygen in your blood. SaO<sub>2</sub> refers to the saturation of oxygen. The value is given as a percentage. The results of this test are not as accurate as the results obtained from an arterial blood gas but they are a good, quick gauge of the oxygenation of your blood. A score of 95-100% is considered normal, but anything over 90% is usually considered adequate. If your saturation dips below 88-90% at rest, supplemental oxygen is usually prescribed.

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\* On a scale of 0-14, seven is neutral. The lower the pH value, the more acid (zero is the most acidic). The higher the pH value, the more alkaline (14 is the most alkaline).

During the test, a small clip with a sensor is placed on one of your fingertips. Within a few seconds, a number shows up on a monitor. It's one of the few tests which are incredibly easy to perform. The only problems are that nail polish or cold fingertips can sometimes give you an inaccurate reading.

You can also have a test called an overnight oximetry to see if you desaturate or lose oxygen while you sleep. Although you may be hooked up to a more extensive monitoring system that will check your heartbeat, pulse and respiration, often a regular oximeter clip will just be taped to one of your fingers. You may even be able to take an oximeter home and perform the test in your own bed.

Many women with LAM have purchased their own pulse oximeters. These are smaller versions than the ones found in most doctors' offices, but they're relatively accurate. Although you don't need to check your oxygen levels constantly, random checks of your levels, especially when you perform different activities like gardening or flying in an airplane, can help your doctor determine if you need supplemental oxygen or if you need a higher liter flow (see the section on Supplemental Oxygen).

### **Six-Minute Walk**

The six-minute walk is a common test used to determine if and/or how much your oxygen levels drop with exertion. Just as it states, a six-minute walk is a test you perform by walking for six minutes, preferably in an area or on a course that has been already measured out in meters or feet. You walk at your own pace to cover as much distance as you can in six minutes. A pulse oximeter is used to measure your heart rate and oxygen level. You'll be asked how tired and how out of breath you are both at the beginning and at the end of the test.

Wear loose-fitting clothing and your gym shoes!

### **Exercise Testing**

One final way to determine the severity of your disease is to perform an exercise test. While you're on a stationary bike or a treadmill, different monitors measure your ventilatory and cardiac limitations, both of which correlate with the severity of your disease. Among other important calculations, the test tells your doctor how much your oxygen saturations drop while you exercise which can be a very important bit of information.

As with the six-minute walk, wear loose-fitting clothes and gym shoes.

## **5.3 Tips for Those Who Need Supplemental Oxygen**

### **General Tips**

- Walk slowly and steadily. You may not need to stop and rest as often.
- Try using a nebulizer if you normally use bronchodilators. You might get an extra boost from this method of delivery.
- Do necessary tasks and harder chores early in the day when you have more energy.
- Rest when you NEED to rest.
- Purchase a pinching/grabbing device for picking stuff up off the floor and for grabbing items down from high shelves. Tasks that require you to bend over or raise your arms will make you more short of breath.

### **Catching Your Breath**

- Sit with your feet on the ground, your elbows on your knees, and lean forward to rest your head in your hands.
- Sit with your feet on the ground, lean forward, and rest your head and your arms on a tabletop.
- Rest your arms on your hips or thighs, or slip your hands into your pockets, or cross your arms to stabilize them.
- Stand with your hands and/or head on a counter or other furniture.
- Relax your neck and shoulders.

### **Nasal Dryness**

- Use a humidifier on your concentrator.
- Try a nonpetroleum-based product in your nose to moisturize it. Some recommended ones are Ocean Saline Spray, Ayr Saline Nasal Gel or Spray, sesame oil, KY Jelly, or Lanisoh (a product for nursing mothers). Don't use: Vaseline, Vicks, or other petroleum products due to risk of lipoid pneumonia from inhaling the tiny petroleum particles into the lungs. Also, petroleum-based products can be flammable.
- Put cocoa butter on your lips to lessen dryness.

### **Showering**

- Use a shower chair in the shower instead of standing.
- Wear a terrycloth bathrobe after bathing to dry yourself without toweling off.
- Wear your cannula in the shower as long as your concentrator is far away.
- Inhale the steam from a nice steamy shower to help you breathe better.

### **Personal Care**

- Find a hairstyle that doesn't require much care. Raising your arms above your head to blow-dry and style your hair will make it harder for you to breathe.
- Sit when you fix your hair and apply make-up. You could bring dressing tables back into fashion.
- Use an electric toothbrush.

### **Equipment**

- Use a conserving device that allows you to carry fewer oxygen tanks with you.
- Switch to liquid oxygen canisters. They are usually smaller than compressed gas canisters and they last longer.
- Connect your hose with a swivel connector so that your hose won't tangle up as easily.
- Understand that although a backpack or shoulder bag for your tank might be easier to get around with, it might be too heavy and you might require a higher liter flow than if you used a pull cart for your tank.

## **Concentrators**

- Pull and stretch your tubing gently to keep it from kinking up.
- Wash your filters weekly.
- Keep a window open, if possible, to add more oxygen to your room for your concentrator to use.

## **On Airplanes**

- Request a seat near the bathroom when you fly with oxygen. You must disconnect your oxygen to leave your seat, so being close to the restrooms will allow you to walk as little as possible without your oxygen.

## **Sexual Intercourse**

- Schedule your intimate encounters during your best breathing time. Do you breathe more easily in the morning, after a midday nap or before bed?
- Experiment to see what position allows you to breathe best. Many women with LAM are more comfortable in upright positions than on their backs.
- Use supplemental oxygen during sexual intercourse, an activity that can be physically draining. If you can enjoy it more and not be exhausted afterwards, you may relax more and indulge in it more often.

## **5.4 Traveling With Supplemental Oxygen**

Try to think positively. Even though traveling with oxygen may be a hassle, you might find yourself more handicapped without oxygen because you are out of breath. Face it: oxygen can allow you more freedom.

With both compressed-gaseous oxygen and liquid oxygen, travel by car doesn't need to be overly complex if you're otherwise healthy. Carefully pack a few tanks in the car and you're off. But NEVER store full tanks in the trunk. The risk of collision from the rear is too great and tanks should NEVER be allowed to roll around! Make sure they are secure. If you'll have access to a source of electricity, small concentrators are available for travel purposes or you can bring your full-size concentrator for lengthier stays in hotels, resorts, or the homes of family and friends.

You can fly with oxygen\* although you can't take your own tanks on board. Generally, the Federal Aviation Administration (FAA) prohibits the use of personal oxygen units during flights because they contain compressed gas or liquid oxygen that is defined as hazardous material. However, the FAA has issued guidelines permitting the onboard use of certain portable oxygen concentrators (POCs). POCs that are approved by the FDA may be carried and used on board most airlines at no charge, in accordance with FAA regulations.

Keep in mind that you are responsible for notifying and making arrangements with the airline(s) which you will be using while you travel. Each airline is different so be sure to go to their website to find out what their specific regulations are. Most airlines require at least 48 hours advance notice but it makes sense to contact them as soon as you book your ticket. Make

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\* If you're on oxygen continuously, you'll definitely need it for flying. If you use oxygen for sleeping and exercising, you may also need it when flying. Ask your doctor if you will need oxygen to fly.

sure that the POC you are using to travel (whether you own the unit or are renting it for your trip) meets the specifications for the airline you are flying with.

Another important factor to consider is the amount of battery power you will need to make your journey. Most airlines require an ample supply of fully charged batteries for at least 150% of your travel time (including layovers on the ground). Each airline is different so be sure to check the airline's website to be sure you are prepared.

Portable POCs are not counted as one of your carry-on items. If you don't need supplemental oxygen on the plane but will need your portable POC at your destination, you are able to carry on your device, as long as it meets the size and weight requirements for carry-on items. They can also be transported as checked baggage. However, keep in mind that batteries that are not being used must be packed separately from the actual POC unit when they are not being used.

Most airlines also require you to have your doctor fill out a form stating why you need oxygen and what liter flow you require. Keep in mind that oxygen for travel, unless it's for your job, may not be reimbursed by your insurance company. Travel is not required for healthy living (even though it may be necessary for your mental health).

The best advice is to always check the airline's policy on supplemental oxygen use. Each airline works a bit differently and rules can change. Even if traveling is a hassle, keep doing it if you enjoy it. Don't forget that the more you can maintain your lifestyle, the better you'll feel mentally, and also physically.



# 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 PaO<sub>2</sub>, 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 PaO<sub>2</sub> 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 (O<sub>2</sub>) is another important factor and is related to the DLCO and the PaO<sub>2</sub>.

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 O<sub>2</sub> 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.

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\* 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

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\*\* 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

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\* 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.

# Chapter Seven:

## Help With the Real World



- 7.1 Social Security and Medicare
- 7.2 Private Disability Insurance
- 7.3 Insurance
- 7.4 Estate Planning and Advanced Directives



## 7.1 Social Security and Medicare

What does it mean to be "disabled"? There are many definitions, so it depends. You might be considered disabled if you can no longer perform the job you were educated or trained to do. If your job is physically demanding, for example, using oxygen 24 hours a day might render you incapable of performing your duties. Some employers may define disability as not being able to perform your duties as you have in the past, while others define disability as being unable to work at any job.

### Social Security Disability

When most people refer to disability benefits, they are referring to Social Security Disability Insurance (SSD or SSDI) rather than a disability plan provided by an employer. If you meet stringent criteria, you may qualify for Social Security Disability benefits.

People who have worked and paid Social Security taxes may be eligible to receive Social Security Disability if they have enough work credits. But how is *disability* defined?

"The definition of disability under Social Security is different than other programs. Social Security pays only for total disability. **No benefits are payable for partial disability or for short-term disability.** Disability under Social Security is based on your inability to work. We consider you disabled under Social Security rules if you cannot do work that you did before and we decide that you cannot adjust to other work because of your medical condition(s). Your disability must also last or be expected to last for at least one year or to result in death."<sup>\*</sup>

The important fact to remember about Social Security Disability benefits is that you must have **recent** work to qualify. Most applicants 31 and older must have 20 credits earned in 5 out of the last 10 years, though younger people may qualify with as few as 6 credits. Benefits tend to change each year with the cost of living, so check with your local Social Security office to find out how much you are eligible for. Or, if you're a worker 25 and older, you can find out how much you're eligible for by reviewing your annual statement from the Social Security Administration that details the amounts you are currently eligible for. The statements arrive each year about two months before your birthday.

Your local Social Security office does not make the medical decision on your disability benefit application. The paperwork is forwarded to a state agency (for example, in Ohio, that is the Disability Determination Service in Columbus), which obtains medical evidence from treating sources. Most cases are decided within 120 days; you may expedite a decision by providing medical records from your physicians and/or hospitals with the initial application. Nationally, about 35 percent of cases are approved at the initial application stage.

If you are found to be disabled, your cash benefits won't start until after a five-month waiting period, which begins the month that Social Security determines you became disabled. If you have limited income and resources, you may qualify for Supplemental Security Income (SSI) during that five-month waiting period (see below). If your application for disability benefits is denied, there are several levels of appeal: reconsideration, hearing before an administrative law judge, Appeals Council review and lawsuit in federal court. If your claim is ultimately approved by an administrative law judge, for example, Social Security will pay you retroactive benefits back to the original filing date or the date you became disabled. For more

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<sup>\*</sup> <http://www.socialsecurity.gov/dibplan/dqualify4.htm>

information or to schedule an appointment, contact your local Social Security Office or call 1-800-772-1213.

You may file for disability benefits over the Internet at the Social Security website, [www.socialsecurity.gov](http://www.socialsecurity.gov). You'll need to print out the application, sign it and submit it to your local Social Security office. You may also complete and electronically transmit the online disability report form (i3368), which asks for information about your medical condition, work history and background. This information is required, and if you don't complete the form online, a Social Security representative will contact you once your electronic application is received.

### **Supplemental Security Income**

If you're 65 or older, or blind or disabled, **and** have little income and few resources, you may qualify for Supplemental Security Income (SSI). While Social Security Disability (SSD) requires you to have worked to be "insured" for benefits, SSI is a needs-based program administered by the Social Security Administration. The definition of disability is the same for both programs. In most states, people who qualify for SSI also qualify for Medicaid, a health-coverage program serving certain eligible low-income people. The program is funded by the federal and state governments and administered at the local level.

### **Medicare**

Medicare, not to be confused with Medicaid, is the federal health-insurance program for people 65 and older, the disabled, and people with end-stage renal disease. A person typically becomes eligible for Medicare after receiving Social Security Disability for 24 months.

Medicare has two parts: Part A, hospital insurance, and Part B, supplemental medical insurance. Part B is optional and requires you to pay a monthly premium that may increase annually. Medicare enrollment is automatic, but you must decline Part B if you don't want it. If you delay taking Part B, you will pay a penalty if you pick it up later, unless you were covered under an employer or your spouse's employer's health plan. For more information about Medicare, visit [www.medicare.gov](http://www.medicare.gov) or [www.cms.gov](http://www.cms.gov).

### **Medicare Drug Coverage after Transplant**

If the costs of your transplant surgery were covered by Medicare (that is, if your transplant was performed at a Medicare-approved facility), Medicare will pay for your immunosuppressant medications after transplant under certain conditions. Check the Medicare website or call your local Social Security office to find out the details.

### **Filing Requirements**

You'll need certain documents to file for SSD, SSI, Medicare and Medicaid. These include your Social Security number, the original or a certified copy of your birth certificate, your latest W-2 or self-employment tax return, your earnings estimate, bank information for direct deposit, information about your marriages and/or divorces, and proof of military service. For SSI, you also must provide information about your income and resources (for example, vehicles, bank accounts, life insurance policies, spouse's income, etc.) to determine whether you meet eligibility requirements.

## 7.2 Private Disability Insurance

If you decide to stop working, find out whether your employer carries private disability insurance. If so, be sure to read the policy. For some workers, private disability insurance may provide an extension or percentage of your current pay and/or benefits.

To a great extent, your enthusiasm for your job and your creativity in solving problems will have a significant effect on your continuing ability to work. If you feel that you're no longer doing your job as well as you'd like, then you should schedule an appointment with your immediate supervisor to discuss your strengths *and* your weaknesses. Adjustments in your work schedule and/or duties might be needed. You may find that you can assume more deskwork (phoning, editing, data processing, etc.) while someone else takes over the more physically demanding work. Ask about new training if you wish to continue working and if the training will allow you to do so. If you and your doctor agree that full-time work isn't good for you, think realistically about part-time work. Keep in mind that your employer probably would prefer to keep you employed and productive in some capacity rather than pay for your disability. Be open to new ideas and new work arrangements.

According to the Americans with Disabilities Act (ADA), employers aren't allowed to discriminate against the disabled in any way, shape or form. A potential employer, however, might be reluctant to hire you if he or she knows you have LAM. If you believe you're being discriminated against because of your illness, you have legal rights to fight that discrimination. Although a lawsuit could be a long and involved process, it's worth your time, effort and money to educate those who discriminate against persons with any form of disability. Don't hesitate to contact a lawyer to help you out if that's the case.

If you plan to file for disability, chances are that your employer will have a procedure spelled out as to exactly what steps need to be taken. Read all of the sections in the employees' manual and then be prepared to ask a lot of questions. When calling to inquire about the process, always write down the date and the name of the person who answered your question. You may be asked to supply that information at a later date.

Once you're on disability, you may discover that there are a limited number of months and/or years that you may receive benefits. If so, you should file for Social Security Disability (SSD) as early as possible because you won't receive SSD benefits until approximately five months after filing.

## 7.3 Insurance

### Private Insurance

If you're disabled, you still need health-care coverage. Larger companies (those with 20 or more employees and those offering a group health plan) are required to offer COBRA (Consolidated Omnibus Budget Reconciliation Act of 1985) for people who stop working. COBRA is a temporary health-care plan that requires your employer to offer you and your dependents whatever health plan you're currently on for the next 18 months, but you must sign up for it within 60 days of leaving your job. The bad news is that your medical insurance costs you more because you must now pay the full amount of the premium; the good news is

that you still have insurance. If you leave work because you're disabled, you qualify for an additional 11 months, bringing the total to 29 months. After 29 months, you're eligible for Medicare.

### ***Affordable Care Act***

The Affordable Care Act was put into place in March 2010 and is aimed at expanding health insurance coverage to Americans who are otherwise not able to afford health insurance. If you are not eligible for Medicaid or Medicare, you may be able to purchase insurance through this lower cost alternative. To see if this option is available to you, visit <https://www.healthcare.gov/>. The website will walk you through the process of finding out if you are eligible, what you qualify for and how to enroll.

### **Life Insurance**

Some life policies state that, if the owner of the policy becomes disabled, he or she doesn't have to pay the premium. Check your policy.

### **HIPAA**

The Health Insurance Portability and Accountability Act of 1997 (HIPAA) is a federal law that was passed to help people keep health coverage if they change jobs, have been laid off or start their own businesses. It was designed to eliminate the possibility that individuals could be denied coverage because of pre-existing medical conditions.

As you might imagine, health insurance companies can get very ingenious in their attempts to "get around" this law. It's important for you to know that you have federal protection in this regard, and it might be in your best interest to get legal help, if necessary, to protect your rights.

### **Long-Term Health Insurance**

This type of policy pays a set amount for a certain time period (it could be for life) if you need to be in a long-term health facility. The policy usually also pays for home health care, therapy, respite care, and a variety of other health-related items, depending on the type of policy you purchase. If you don't have such a policy before you're diagnosed with a long-term illness like LAM, you may have trouble qualifying for a new policy of this nature.

### **Other Insurance**

Life insurance, car insurance, homeowner's insurance, single-disease insurance, airline insurance, and so on—where does it stop? So many policies are available. Discuss with your primary insurance agent whether any fit your needs and situation.

### **Getting Free Help**

Advocating for Chronic Conditions, Entitlements, and Social Services (ACCESS) is a free service offered by Olsten Health Services, a privately run agency. ACCESS can help you with Social Security Disability, Medicare, Medicaid, and Supplemental Security Income. It can also help those who are still working hang on to their health insurance under COBRA or HIPAA. Call 1-888-700-7010. The Department of Labor will also send you a publication on recent changes in health-care laws. Call 1-800-998-7542.

## **7.4 Estate Planning and Advanced Directives**

### **Estate Planning**

Writing a will should be a required task for any person, not just for a woman with LAM. Your will should make provisions for your children and their care and for the distribution of your assets. Not only will getting your affairs in order give you peace of mind, but your heirs will be relieved by your advanced planning.

You can actually write a will yourself using a form from an office-supply store or simply by going online, but most people feel more comfortable obtaining professional legal services. Ask your attorney up front what the charges will be. If the fees are too expensive, call another lawyer. When you find a lawyer you trust and who fits your budget, don't be afraid to ask questions or to specify exactly what you want in the will. It's your will and the decisions are yours. When you are providing for your children and distributing your estate, don't neglect charitable organizations if you have sufficient funds. If you would like to list The LAM Foundation as one of your beneficiaries, contact the Foundation for more information.

If you are concerned about having sufficient funds to take care of your future expenses, you might feel it necessary to consult a financial planner. A financial advisor can help you make your money work for you so that you have adequate monies to live on. Once again, don't be afraid to ask questions including what the financial advisor's fee will be.

Finally, some people choose to prepay their funeral expenses to save their loved ones the stress of making all the arrangements during a time of grieving. Most funeral homes offer this service, and many churches have files where their members can record their wishes about final services. Again, you should look at several options since funeral costs can vary significantly.

### **Advanced Directives**

Don't be squeamish about preparing an advanced directive. These types of documents are as important as any will or any estate planning that you do. The National Institutes of Health regularly advises each of its patients enrolled in the LAM protocol to have an advanced directive on file and many transplant centers require one for each prospective transplant patient. Advanced directives designate a competent person to act as your agent in matters like health care and finances should you be unable to do so yourself. These documents not only insure that your wishes will be carried out but they also make decision-making easier for your loved ones. Think of advanced directives as insurance policies, only slightly different from life insurance or disability insurance. There are three main types of advanced directives: living wills, durable powers of attorney for health care and durable powers of attorney.

### **Living Wills**

A living will is a document that should be considered by all people regardless of their medical conditions. This document allows you to direct your future medical care should you be unable to make your wishes known. If you should become incapacitated and can't tell the medical experts what you wish them to do, a living will would make certain that your wishes would be followed.

Living wills are generally available from attorneys, physicians, hospitals, bar associations, nursing homes and some nonprofit organizations. Nowadays, many hospitals ask you for one when you are admitted or if you are preparing to be hospitalized. Generally, the hospital staff

has forms for you to fill out and sign so that, should something unforeseen occur and should you lose your ability to make decisions, your wishes would be known. If you sign one when you are admitted and change your mind later, you may revoke your living will at any time.

Although many people have living wills, frequently the will is not readily available at the time decisions need to be made. Therefore, to insure that your wishes are carried out, keep the original document wherever you keep all of your important papers. But first, make at least seven copies of your living will and other advanced directives. Give one copy to your spouse or to a family member, one to your best friend, one to your doctor to keep with your medical records and one to your lawyer. Also, keep a copy in your nightstand, a copy in your safety deposit box and a copy in the glove compartment of your car.

### **Durable Power of Attorney for Health Care**

A durable power of attorney for health care allows you to name a person (your spouse, relative, friend or your attorney) to act as your attorney-in-fact to make health-care decisions for you. There is a difference between a living will and a durable power of attorney for health care: a living will makes your wishes known directly to your doctors concerning medical care, especially life support; a durable power of attorney for health care allows for someone you have chosen and who knows your wishes, to make decisions regarding your medical care if you can't act for yourself. If there is a disagreement between your family and the doctor treating you, your *written* wishes can be the deciding factor. Just like a living will, you can revoke this document at any time. When writing a durable power of attorney for health care, be sure to include your wishes to be an organ donor!

### **Durable Power of Attorney**

A durable power of attorney provides someone you designate broad powers to handle your property. The durable provision enables your agent to carry out your wishes should you become incompetent. This document does not authorize anyone to make medical or other health-care decisions, but it would allow your agent to make other decisions such as the care of your children if you have not made provisions for them or the disbursement of funds if you are unable to make financial decisions. This document is routinely written during the process of writing a will. Legal forms are available in office-supply stores, online or from your family lawyer.

### **Organ Donation**

If you decide to donate your organs or tissue, you should include your wishes in your advanced directives. But you *must* tell your family members of your wishes. Even if you have your desire to be an organ donor in writing in your living will, in your durable power of attorney for health care, or on your driver's license or some other document, your family may still be asked to give their consent before you can become a donor. Be sure to tell each and every family member of your wish to donate.

# Chapter Eight:

## Odds and Ends



- 8.1 About The LAM Foundation
- 8.2 Alphabet Soup
- 8.3 Glossary of Terms
- 8.4 Sources of Information
- 8.5 Leading a Hopeful Life



## **8.1 About The LAM Foundation**

### **Our Mission**

The LAM Foundation urgently seeks safe and effective treatments, and ultimately a cure, for lymphangioleiomyomatosis (LAM) through advocacy and the funding of promising research.

We are dedicated to serving the scientific, medical and patient communities by offering information, resources and a worldwide network of hope and support.

### **Our History**

The LAM Foundation is the global leader in the fight against lymphangioleiomyomatosis (LAM). Founded in 1995 as a grass roots effort, The LAM Foundation has evolved into an organization that is described by the National Heart, Lung and Blood Institute (NHLBI) as "a model for voluntary health agencies."

Headquartered in Cincinnati, Ohio, The LAM Foundation provides support and education for women with LAM and their families, engages doctors and scientists to continue to learn more about the disease and raises funds for the continued study of LAM.

The Foundation has raised and invested more than \$25 million in research over the last 24 years, resulting in the fundamental understanding of the genetic cause of LAM which led to the first ever clinical treatment trial and ultimately an FDA approved treatment for the disease.

More than 2,000 women with LAM have been identified to date but it's suspected that there may be as many as 250,000 undiagnosed or misdiagnosed living with LAM. The LAM Foundation strives to provide hope for all women with LAM by providing a network of support and continued research to ultimately find a cure for LAM.

### **Patient Services**

As a part of our dedication to the LAM patient community, The LAM Foundation strives to be the premier resource for all LAM patients. The following programs are just a few of the services available to help women with LAM receive the best care and support possible.

#### *LAM Liaison Patient Network*

To help women with LAM connect with one another, the Foundation developed the LAM Liaison Patient Network. This network was created by dividing the United States into 21 regions. Regions were determined based on patient numbers, geography and proximity to an established and/or future LAM Clinic. To see a map of the regions, visit our website (<https://www.thelamfoundation.org/Newly-Diagnosed/More-Resources/LAM-Liaison-Patient-Network>).

A LAM Liaison is a regional leader who reaches out to patients in her region and also organizes one or more regional meetings each year. These meetings will give you the opportunity to meet other patients who live in your area and discuss issues that are important to you. You are welcome to attend meetings in any region. Family and friends are also encouraged to attend regional meetings. Visit our website to review the scheduled regional meetings taking place across the United States.

The LAM Liaison Patient Network ensures that all women with LAM receive the personal attention they both need and deserve. Contact your LAM Liaison and/or Area Lead with any questions you might have about living with LAM.

### *Publications*

The LAM Foundation strives to keep our community up to date on the latest information about research and what is happening with other members of the community. Our monthly e-newsletter called Currents is emailed out on the last day of each month. This is full of up-to-date information about events and accomplishments. Journeys is our magazine publication that is mailed out to patients, family members, donors, doctors and scientists twice each year. This publication gives us an opportunity to provide in-depth information to all who are touched by LAM.

### *The LAM Foundation Website*

The LAM Foundation website (<https://www.thelamfoundation.org>) is the premier source of information about lymphangiomyomatosis. It was put together to serve several distinct users, including newly diagnosed patients, patients living with the disease, family members and caregivers of LAM patients, healthcare providers caring for LAM patients and scientists who are interested in doing research about LAM.

## 8.2 Alphabet Soup

ABG	arterial blood gas
ACCESS	advocating for chronic conditions, entitlements, and social services
ADA	Americans with Disabilities act
AML	angiomyolipoma
ATS	American Thoracic Society
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BAL	bronchial alveolar lavage
BID	twice daily (latin: <i>bis in die</i> )
BMD	bone mineral density
BP	blood pressure
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CAT	computerized axial tomography
CMV	cytomegalovirus
CO	carbon monoxide
CO <sub>2</sub>	carbon dioxide
COBRA	consolidated omnibus budget reconciliation act
COPD	chronic obstructive pulmonary disease
CPFR	certified pulmonary function technician
CT	computerized tomography
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DLCO	diffusing capacity for carbon monoxide
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ECG	electrocardiogram
EKC	electrocardiogram
ER	emergency room
ERV	expiratory reserve volume
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FEF25-75%	average forced expiratory flow over the middle half of the fvc
FEFMax	forced expiratory flow maximum
FET	forced expiratory time
FEV1	forced expiratory volume in one second
FEV1/FVC	the percent of the vital capacity that can be forcibly expelled in the first second
FRV	functional residual capacity
FVC	forced vital capacity
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HIPAA	health insurance portability and accountability act
HMO	health maintenance organization
HR	heart rate
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IC	inspiratory capacity
IRV	inspiratory reserve volume
IU	international unit
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LAM	lymphangiomyomatosis
LPM	liters per minute
MDI	metered-dose inhaler
MMEF	maximum mid expiratory flow
MRI	magnetic resonance imaging
NHLBI	National Heart, Lung, And Blood Institute
NIH	National Institutes Of Health
NKDA	no known drug allergy
NKFA	no known food allergy
NORD	National Organization Of Rare Diseases
NP	nurse practitioner
NPO	nothing by mouth (latin: <i>nils per os</i> )
NTAF	national transplant assistance fund
O <sub>2</sub>	oxygen
O <sub>2</sub> sats	oxygen saturations
OTC	over the counter
PA	physician's assistant
PACO <sub>2</sub>	arterial partial pressure of carbon dioxide
PAO <sub>2</sub>	arterial partial pressure of oxygen
PEF	peak expiratory flow
PEFR	peak expiratory flow rate
PES	post embolization syndrome
PF	peak flow
PFTs	pulmonary function tests
PO <sub>2</sub> or PaO <sub>2</sub>	partial pressure of oxygen
PCO or PaCO <sub>2</sub>	partial pressure of carbon dioxide
PRN	take medication as needed (latin: <i>pro re nata</i> )
PTX	pneumothorax
QD	once a day (latin: <i>quaque die</i> )
QID	four times a day (latin: <i>quarter in die</i> )
RPFT	registered pulmonary function technician
RRT	registered respiratory therapist
RV	residual volume
SOB	short of breath
SSD	social security disability
SSI	social security insurance

TID	three times daily (latin: <i>ter in die</i> )
TLC	total lung capacity
TPN	total parenteral nutrition
TS	Tuberous Sclerosis
TSC	Tuberous Sclerosis Complex
TV	tidal volume

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UNOS	United Network For Organ Sharing
URI	upper respiratory infection

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VATS	video-assisted thoracic surgery
VC	vital capacity

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### 8.3 Glossary of Terms

**advance directive** — An advance directive tells your doctor what kind of care you would like to have if you become unable to make medical decisions (if, for example, you're in a coma).

**air-trapping** — The retention of a significant amount of air in one's lungs, even after both normal and maximal exhalation.

**alveoli** — Tiny air sacs at the ends of the bronchial tubes in the lungs where gas exchange occurs. The transfer of carbon dioxide from the blood into the lungs and of oxygen from the air into the blood takes place in the alveoli.

**Angiography** — The examination of blood vessels by using x-rays after the vessels have been injected with a radiopaque substance.

**angiomyolipoma (AML)** — A benign tumor (oma) consisting of blood vessels (angio), muscles (myo), and fat (lipo). AMLs occur in TS and in LAM and are most often found in the kidneys.

**arterial blood gas (ABG)** — A measurement of the oxygen and carbon monoxide in the blood taken by drawing a blood sample from an artery (usually the radial artery near one's wrist or in the brachial artery in the crook of the arm) rather than a vein. This measurement is considered the most valid in determining the amount of oxygen in the bloodstream.

**aspiration** — 1. The use of a small needle or catheter to suction fluid from a cavity of the body.  
2. The inhalation of fluid or a foreign substance, such as vomit, into the bronchi and lungs.

**barium** — A metallic element. Barium is mixed with certain drinks and is taken before some x-ray procedures to visually enhance the image taken.

**bilateral transplant** — Receiving two lungs during a transplant. Also called a double-lung transplant.

**biopsy** — A small sample of tissue taken from an organism for diagnostic purposes.

**bleb** — A cyst full of fluid and/or air on or near the surface of the lungs.

**bronchial alveolar lavage (BAL)** — The introduction of a small amount of a sterile saline solution into the lungs during a bronchoscopy to "wash out" tissue samples for study.

**bronchitis** — A disease characterized by inflammation of the mucous membranes of the bronchial tubes.

**bronchodilator** — An agent/medicine that relaxes and enlarges the opening of a bronchus or bronchial tube.

**bronchodilator response** — An expansion of the bronchial tube, after using an agent/medication that allows improved air flow. An example of a bronchodilator response is when your PFT results are better after you inhale a medication like Albuterol.

**bronchoscopy** — A procedure using a flexible fiberoptic tube (bronchoscope) to examine the interior of the lungs.

**bronchus** — *pl. bronchi* One of two branches leading out of the trachea and into the lungs.

**bullae** — A bleb.

**capillary** — A tiny blood vessel.

**cardio-pulmonary exercise test** — An exercise test performed on a treadmill or exercycle designed to test exercise capacity by measuring oxygen saturation, pulse, blood pressure, and heart rate.

**chest cavity** — The space in the body surrounding the lungs.

**chest tubes** — Sterile tubes inserted into the chest cavity between the lung and chest wall to reinflate the lung or to release air or to drain chyle or another fluid from the area.

**Chronic Obstructive Pulmonary Disease (COPD)** — An obstructive lung disease most frequently caused by smoking. Although LAM is an obstructive lung disease also, most physicians don't refer to it as COPD.

**chyle** — A lymphatic fluid that drains from the lymph glands. It is milky in color and contains fat and other nutrients.

**chyloptysis** — Expecterating (spitting up/coughing up) chyle.

**chylothorax** — Chyle in the chest cavity around the lungs that obstructs the efficient function of the lungs.

**chylous ascites** — The abnormal accumulation of chyle in the abdominal cavity.

**chylous effusion** — The abnormal accumulation of chyle in the chest cavity/pleural space.

**chyluria** — Passage of chyle into the urine.

**collapsed lung** — A condition where the space between the chest wall and the lung is filled with air or fluid causing the lung to deflate partially or completely. Also called a pneumo or a pneumothorax.

**Computerized Tomography Scan (CT or CAT=computerized axial tomography)** — An x-ray process where a machine takes pictures of “slices” or planes of the body at specified intervals. These images give the impression of looking into the body from the top instead of from the front, back, or side. CT scans look more three-dimensional than standard x-rays.

**cyanosis** — A bluish tint or discoloration of the skin and/or mucous membranes due to insufficient oxygenation of the blood.

**CytoMegalovirus (CMV)** — A herpes-type virus. Every person is either CMV positive or negative. Most of the population is positive, meaning they have been exposed to the virus and have developed antibodies for it. People who are negative have no antibodies for this virus. CMV infects the salivary glands. Symptoms, if there are any, are like mononucleosis. CMV can cause serious illness if a person is immunosuppressed, for example, after transplant.

**Depo Provera** — A drug, administered by injection, that contains a synthetic hormone similar to the natural hormone progesterone.

**Diffusing Capacity for Carbon Monoxide (DLCO)** — The use of carbon monoxide to measure the diffusion capacity (how well the gasses pass through the membranes) of the lung.

**dyspnea** — Shortness of breath (SOB) or difficulty breathing.

**echocardiogram** — The use of ultrasound to examine the heart.

**electrocardiogram (EKG or ECG)** — This test is used to measure the activity of the heart muscle. Electrodes are placed on the chest and extremities, and an electrical current, which passes through the body, is used to detect abnormalities in the heart.

**embolization** — Therapeutic introduction of various substances into a vessel to occlude or block it. The procedure is used either to arrest or prevent hemorrhaging or to cut off the blood supply to a structure or organ, to destroy or weaken it.

**endotracheal** — Inside of or going through the trachea.

**etiology** — The cause of a disease.

**exertional dyspnea** — Shortness of breath on exertion.

**Expiratory Reserve Volume (ERV)** — The extra amount of air you can forcibly exhale after you’ve exhaled normally.

**FEV1/FVC** — The percent of the vital capacity that can be forcibly expelled in the first second.

**fibroma** — Benign tumors consisting primarily of fibrous tissue.

**flame** — To express your opinion online in a very strong, heated, emotional, and generally disagreeable manner.

**Forced Expiratory Time (FET)** — The amount of time (in seconds) the patient exhales during the FVC test.

**Forced Expiratory Volume in One Second (FEV1)** — The volume of air (in liters) one can forcibly exhale in one second.

**Forced Vital Capacity (FVC)** — The total volume of air (in liters) expelled after a full inhalation.

**hemoptysis** — Coughing or spitting up blood or blood-stained sputum from the lungs or the bronchial tubes.

**hypoxemia** — Below normal amount of oxygen in arterial blood; lack of sufficient oxygenation of the blood.

**hypoxia** — A condition in which you and your tissues are oxygen deprived.

**Inspiratory Capacity (IC)** — The total amount of air that you can draw into your lungs after a normal exhalation.

**Inspiratory Reserve Volume (IRV)** — The amount of air that you can take in from a normal resting inspiration to the fullest inspiration.

**intubation** — Placing a tube into trachea.

**LAMposium** — An annual conference to educate and support LAM patients, their families, and the medical/scientific personnel interested and involved in LAM research.

**larynx** — Voice box.

**leiomyo** — Smooth muscle.

**leiomyoma** — A benign tumor of smooth muscle. In the uterus, a leiomyoma is known as a fibroid.

**leiomyomatosis** — The state of having multiple *leiomyomas*.

**lipid-free diet** — A restrictive diet to decrease fat intake.

**Listserv** — An e-mail discussion site set up to provide support, information, and fellowship to e-mail users who share a common cause.

**lurking** — Monitoring e-mail messages on a Listserv before joining in the discussion.

**lymph** — A clear, slightly yellow fluid, containing white blood cells, found in the lymphatic vessels. Lymph is collected from tissue fluids throughout the body and returned to the blood via the lymphatic system.

**Lupron** — An anti-estrogen drug which is injected and which is often used to treat prostate cancer in men.

**Magnetic Resonance Imaging (MRI)** — A sophisticated imaging scan using magnetism to disrupt cells temporarily and then monitor the energy the cells produce as they return to their normal state. MRIs are often used for internal structures of the body, especially, but not restricted to, soft tissues and the brain.

**metered-dose inhaler** — A device that holds a little aerosol metal canister of medicine to be inhaled. This device releases the exact dose of medicine in small “puffs.”

**national registry** — A national database built from information about a group with a commonalities. Information about diseases is often gathered and made accessible to those studying the particular disease through databases.

**nebulizer** — A device that is pressurized by an oxygen tank and that converts a liquid medicine into a fine mist that can be inhaled. (This is the device you breathe from during your PFTs when you’re getting the Albuterol.) Nebulizers are often used in hospitals or when you are unable to use a metered-dose inhaler.

**nephrectomy** — Removal of a kidney.

**obstructive lung disease** — A lung disease in which the person experiences limited airflow.

**osteopenia** — A decrease in the density of bone mass, often leading to osteoporosis.

**osteoporosis** — A disease in which the bones lose density and become very porous.

**oxygen (O<sub>2</sub>)** — Two molecules of oxygen. The form of oxygen that is in the air we breathe.

**oxygen (supplemental)** — Oxygen received through a nasal cannula or mask to raise oxygen levels in the blood.

**oxygen saturation (O<sub>2</sub> sats)** — The percentage of oxygen measured in your blood. Oxygen saturations are usually measured (totally painlessly) by wearing a small contoured clip called a pulse oximeter on the tip of your finger for a minute or two. Ideal oxygen saturations are above 90%.

**oximeter** — See **pulse oximeter**.

**pericardial effusion** — An increased amount of fluid in the sac around the heart.

**pleura** — A thin membrane that covers the outside of each lung and folds back to make a lining for the inside of the chest cavity.

**pleural abrasion** — Roughing up of the pleura by one of various agents to attach the lung to the inside of the chest cavity. Pleural abrasion is used in attempts to repair and prevent lung collapses.

**pleural effusion** — A collection of fluid in the pleural space (chest cavity).

**pleurectomy** — The removal of the pleura, the membrane enveloping the lungs and lining the walls of the chest cavity, usually performed to adhere the lungs to the walls of the chest to keep the lungs inflated.

**pleurodesis** — A procedure to adhere of the lining of the lung to the lining of the chest cavity. The procedure eliminates the space between the lungs and the chest wall and prevents further lung collapses.

**pleuroperitoneal shunt** — A small tube placed inside the body that connects the chest cavity to the abdominal cavity to drain fluid from the chest.

**pneumoretropharynx** — A condition that occurs after a pleurodesis when air travels up the airway from the lungs to the back of the throat, causing a "pain in the neck."

**pneumothorax** — A lung collapse. "Pneumo" refers to air and "thorax" to the chest cavity. The air leaks into the space between the lining of the lung and the lining of the chest. The pressure of the air between the lung and the chest wall forces the lung to collapse.

**predicted value** — The averaged scores of a reference population to which your test results are compared.

**Pulmonary Function Tests (PFTs)** — A series of test to determine how well the lungs function by checking the performance of the lungs and determining the severity of obstruction in the airways.

**pulmonary hemorrhage** — Bleeding into or from the lung.

**pulmonary rehabilitation** — A program of exercise and education to increase pulmonary function and to strengthen the body so it can function more efficiently.

**pulse oximeter** — A small device that clips onto a fingertip and gives a reading of the oxygen concentration in the blood at that particular moment.

**pneumovax** — A vaccination to prevent pneumonia.

**PO<sub>2</sub> or PaO<sub>2</sub>** — **Partial pressure of oxygen.**

**PCO or PaCO<sub>2</sub>** — **Partial pressure of carbon dioxide.**

**renal AML** — An angiomyolipoma in the kidney.

**Residual Volume (RV)** — The volume of air remaining in the lungs after maximal exhalation.

**restrictive lung disease** — A lung disease in which airflow is restricted both in and out of the lungs.

**smooth muscle cell proliferation** — The growth and reproduction of smooth muscle cells. In LAM, these cells take up space meant for lung tissue and form the “blebs” or “cysts” in the lungs.

**spacer** — A large plastic container with a mouthpiece at one end and a hole at the other end for inserting the mouthpiece of a metered-dose inhaler. A spacer makes it easier to deliver the medication directly into the lungs and not into the mouth and throat, reducing side effects.

**spirometer** — A computerized machine that measures the volume of air and the speed of exhalation.

**spirometry** — A series of tests that are performed on a spirometer and that measure the air flow into and out of your lungs.

**Sporadic LAM** — A type of LAM differentiated from LAM that occurs in conjunction with Tuberous Sclerosis. In Sporadic LAM, the genetic mutations occur only in the LAM cells of the organs that are affected, not in the reproductive cells.

**SOB-ness** — Short of breath-ness. The condition of being short of breath (coined by a LAM patient).

**talc poudrage** — Pleurodesis using medical grade, sterile talc powder blown into the pleural space.

**talc slurry** — Pleurodesis using medical grade, sterile talc liquid injected into the pleural space.

**thoroscopic surgery** — Surgery performed inside the chest cavity using a thoroscope (an optical instrument with a lighted tip) to assist the surgery. The technique is also called VATS (video-assisted thoracic surgery).

**thoracotomy** — Open-chest surgery; an incision into the chest wall for a biopsy or for a treatment.

**thoracentesis** — The passage of a hollow tube or needle into the chest cavity to draw off fluid.

**Tidal Volume (TV)** — The amount of air you inhale and exhale during your resting or normal breathing pattern.

**Total Lung Capacity (TLC)** — The total volume of air in the lungs after a maximal inspiration.

**trachea** — The tube connecting your mouth and nose to your lungs; windpipe.

**transplant** — The transfer of an organ (or tissue) from one body to another.

**TS-related LAM** — LAM that occurs in conjunction with Tuberous Sclerosis.

**Tuberous Sclerosis (TS) or Tuberous Sclerosis Complex (TSC)** — A genetic disorder characterized by tumors of the brain, heart, kidney, lungs and skin; seizures; and mental retardation. Not all individuals with TSC will have all of these symptoms.

**twenty-four hour urine** — A test during which all the urine produced during a 24-hour period is saved and tested for the presence of certain chemicals cleared from the body by the kidneys. It provides information on the function of the kidneys.

**ultrasonography** — See **ultrasound**.

**ultrasound** — A way of viewing internal organs using sound. A handheld “microphone” is moved over the area of an organ to detect abnormalities.

**unilateral transplant** — A single-lung transplant.

**Video-assisted thoracic surgery (VATS)** — Surgery using an optical instrument with a lighted tip. (See **thoroscopic surgery**.)

**Vital Capacity (VC)** — The amount of air that you can forcibly exhale after a full inspiration.

**windpipe** — Trachea.

## 8.4 Sources of Information

The LAM Foundation

Website: <https://www.thelamfoundation.org> Phone: 877-287-3526

National Institutes of Health

Website: <http://www.nih.gov> Phone: 877-644-5864

MedicAlert® Medical-Identification Jewelry

Website: <http://medicalert.com> Phone: 888-633-4298 (toll-free)

Lauren’s Hope Medical-Identification Jewelry

Website: <http://www.laurenshope.com> Phone: 800-360-8680

TS Alliance

Website: <http://www.TSAlliance.org> Phone: 800- 225-6872

Social Security Administration

Website: <http://www.socialsecurity.gov> Phone: 800-772-1213

Medicare

Websites: <http://www.medicare.gov>, <http://www.cms.gov>

ACCESS (Advocating for Chronic Conditions, Entitlements, and Social Services, for help with SSI, SSD, Medicare and Medicaid)

Phone: 888-700-7010.

The Department of Labor

Phone: 800-998-7542

#### References for Traveling with Oxygen

Breathin' Easy; A Guide for Travelers with Pulmonary Disabilities

Website: <http://oxygen4travel.com>

#### Transplantation - Helpful Websites

Medline Plus, a service of the U.S. National Library of Medicine and the National Institutes of Health

Website: <http://www.nlm.nih.gov/medlineplus/lungtransplantation.html>

Duke Transplant Center

Website: <http://organtransplant.mc.duke.edu/transplant.nsf>

Cleveland Clinic Lung Transplant Program

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

Transplant Living

Website: <http://www.transplantliving.org> (from the *Before the Transplant* menu, click on *Financing a Transplant*)

United Network for Organ Sharing (UNOS)

Website: <http://www.unos.org> Phone: 888-TXINFO1

Second Wind (the national support organization for lung transplantation)

Website: <http://www.2ndwind.org>

HelpHopeLive

<https://helphopelive.org/>

## 8.5 Living a Hopeful Life

To learn to cope with LAM, you need to become aware of the power you hold within you: the power of hope! You must choose to hope. Why? Because patients who have hope tend to live longer and have a better quality of life. For them, hope acts as a medicine.

Where do you find hope? For some, hope is found in family ties, books or the wonders of modern medicine. For others, it's found in a deep spirituality, a belief in God, in a universal power or in one's own inner strength. It may take you a while to find hope, but you can find it.

Having hope softens the little and the big setbacks, and it puts you in control. You still feel pain and grief, but hope gives them a different resonance and gives you more strength to face the future. It's much harder to be hopeful when you're feeling sick and mad at the world. And sometimes, when things are really, really bad, you may have no hope at all. But you can find hope again if you search for it.

What should you hope for? Anything you want. Some women hope for a cure for LAM. Some hope to live a long and happy life and to see their children and grandchildren grow up. And others simply hope to be at peace with whatever each day brings. Your positive attitude can allow you accept your diagnosis and move on with your life. It's up to you to choose how you react to LAM and its problems. But never let your diagnosis, the progress of your disease or the sad statistics take hope away from you.

To aid you on your journey, here are two small, but hope-filled, sentiments.

### Native American Blessing

Hold on to what is good, even if it is only a handful of earth.  
Hold on to what you believe, even if it is a tree that stands by itself.  
Hold on to what you must do, even if it is a long way from here.  
Hold on to life, even if it is easier to let go,  
Hold on to my hand, even when I have gone away from you.

### Hope Is the Thing with Feathers

Hope is the thing with feathers  
That perches in the soul  
And sings a tune without words  
And never stops at all.

And sweetest, in the gale, is heard  
And sore must be the storm  
That could abash the little bird  
That keeps so many warm.

I've heard it in the chilliest land  
And on the strangest sea  
Yet, never, in extremity  
It asked a crumb of me.

— Emily Dickinson

