

MESOTHELIOMA

A Patient's Road Map

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Established in 2002, the Pacific Heart, Lung & Blood Institute ("PHLBI") is a 501(c)(3) non-profit institution focused on the treatment and prevention of malignant pleural mesothelioma. The Institute supports the research efforts of renowned mesothelioma surgeon Dr. Robert Cameron, who advocates the lung-sparing Pleurectomy/Decortication surgery for patients diagnosed with malignant pleural mesothelioma.

Learn more about mesothelioma treatment options, Dr. Cameron's innovative approach to treating mesothelioma and the research which may someday lead to a cure by visiting the Institute's website www.phlbi.org.

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Mesothelioma: A Patient's Roadmap

*Presented by
the Pacific Heart, Lung & Blood Institute*

Patients diagnosed with mesothelioma find themselves at the beginning of a journey. Because of the rarity of the disease and the developing nature of the treatments, it is a journey that will take them on sparsely-traveled, out of the way back-roads. The kind of roads that are often difficult to navigate. Difficult, that is, unless you have a good roadmap.

The Pacific Heart, Lung & Blood Institute has assembled this Patient's Roadmap to help mesothelioma patients navigate the many roads before them on their journey to proper diagnosis, treatment and coping with their disease. With this map, mesothelioma patients can more quickly and efficiently find their way.

But, for those who think they may be lost, additional information or "Roadside Assistance" can be obtained by visiting the Institute's website **www.phlbi.org** or contacting the Institute at **(310) 478-4678** or **info@phlbi.org**.

What is Mesothelioma?

Mesothelioma is an aggressive form of cancer which is diagnosed in about 2,000 to 3,000 people in the United States every year. The disease gets its name because it initially affects the mesothelium, a protective membrane that covers internal organs of the body including the lungs, heart and abdominal organs. Mesothelioma can develop at any of these sites, but approximately 75% of the time it is the lung lining, or pleura, that is affected.

The only known cause of mesothelioma is exposure to asbestos. Asbestos is the general name given to a group of minerals which can be separated into durable threads or fibers. Because it is resistant to heat, fire, electricity and chemicals, asbestos was used in thousands of industrial, construction, automotive and commercial products through the years.

Unfortunately, the indestructible properties which make asbestos so effective in these products are also what make asbestos extremely toxic to the human body.

Asbestos fibers are finer than human hair and invisible. The fibers are easily inhaled and, because of their long thin shape, are often immune to the body's natural defense mechanisms against foreign bodies entering the airways. The fibers are able to proceed past the lung and lodge into the pleura.

Over the course of 20 to 60 years, the fibers can cause inflammation and fibrosis which damage the DNA of the mesothelial cells lining the pleura and lead to the appearance of malignant tumors. If left untreated, the tumors can spread throughout the pleural space as a sheet of fibrous tissue which surrounds the lung, exerting increasing pressure on the lungs and surrounding organs.



How is Mesothelioma Diagnosed?

Proper diagnosis of mesothelioma requires a combination of physical, radiological and pathological examinations, as well as the taking of an accurate history including potential asbestos exposure.

The most common physical symptoms of mesothelioma are shortness of breath, chest pain, unproductive cough, weight loss and loss of energy.

Because these symptoms are common to many illnesses, mesothelioma often goes undetected as doctors look to more common causes of the symptoms. For this reason, it is important to raise the doctor's level of suspicion for mesothelioma by making sure that the history of asbestos exposure is revealed in the admission questionnaire or during the history taken by the doctor or nurse.

Presented with the combination of these symptoms and a history of asbestos exposure, doctors will frequently order radiological testing such as an X-ray, CT scan, PET scan or MRI. Abnormalities revealed in any of these studies, such as a pleural effusion (excess fluid in the pleural linings of the lungs) or a suspected mass, should prompt additional testing.

For patients with a pleural effusion, a thoracentesis (drainage of the fluid) will typically be performed to ease the pressure exerted on internal organs and reduce discomfort. A sample of the fluid will be examined by a pathologist for malignant cells. However, given the high rate of "false negative" fluid cytology results for mesothelioma, experienced doctors will typically proceed with a tissue biopsy even if the fluid cytology is negative.

The only reliable test for diagnosing mesothelioma is a tissue biopsy, or removal of a small piece of tissue for laboratory examination. There are several different types of biopsies.

A **needle biopsy** involves passing a needle through a syringe into the

area of concern, often with the aid of an x-ray or CT scan, and removing tissue using the needle. An **open biopsy** is a surgery performed under general anesthesia involving an incision into the affected area and removal of tissue. A **closed biopsy** involves a much smaller surgical incision and use of a small camera-like instrument which guides the surgeon to the appropriate place to take the sample.

Examination of the biopsy samples includes immunohistochemical (IH) staining in which the pathologist looks for specific signs, or markers, of mesothelioma. This is often a complex process which requires analysis by a pathologist who is experienced in diagnosing mesothelioma and the specific cell types of the disease (epithelial, sarcomatoid, desmoplastic, biphasic). It is not uncommon even for experienced pathologists to seek a second opinion when diagnosing mesothelioma.

What is Staging and Why is it Important?

Once a diagnosis of malignant mesothelioma is reached, more tests will be performed to determine whether cancer cells have spread to other parts of the body. This is called "staging". Doctors will need to know the stage of the cancer in order to determine available treatment options.

When staging mesothelioma, doctors investigate where the cancer is located, the size of the tumor, whether the cancer has spread to other body parts, and whether the cancer is affecting other bodily functions. The standard for staging pleural mesothelioma is as follows:

Stage I: The cancer is located in the pleura and has not spread to the lymph nodes or other organs. Stage IA refers to a tumor in the outer layer of the pleura, whereas Stage IB refers to a tumor that has reached both the outer and inner layer.

Stage II: The cancer has spread beyond the pleura and into the lung tissue and/or diaphragm, but not yet into the lymph nodes.

Stage III: The cancer has spread into the fatty part of the mediastinum (the space between the lungs and behind the breastbone) and/or into the lymph nodes within the thorax.

Stage IV: The cancer has spread to other organs such as the heart, opposite lung, peritoneum or spine.



What are the Treatment Options for Mesothelioma?

As with other forms of cancer, the goal in treating mesothelioma is to remove or destroy malignant cells while preserving healthy cells. At the present time, the three most frequently used options for treating mesothelioma are surgery, chemotherapy and radiation. There are also a number of newer or experimental treatments which are showing promise in the treatment of mesothelioma. Because of certain limitations associated with each of these treatments, they are often used in differing combinations by mesothelioma specialists.

I. Surgery

The purpose of surgery is to remove all visible tumor, thereby relieving pressure which the growing tumor places on the internal organs and preventing the tumor from spreading to other parts of the body. While, in a successful surgery, all of the tumor is removed, some amount of cancer cells will inevitably remain. For this reason, surgery is almost always used in combination with another treatment to kill the remaining cancer cells.

There are two surgical procedures which are used for treating mesothelioma: the Extrapleural Pneumonectomy and the Pleurectomy/Decortication.

The **Extrapleural Pneumonectomy**, or "EPP", is a radical procedure which involves the surgical removal of the entire lung, a portion of the diaphragm (the muscle located below the lung) and the pericardium (the lining of the heart). Because of the radical nature of this procedure, and the fact that the patient will be left post-operatively with only one lung, it can only be performed on patients who meet certain criteria such as good cardiac and pulmonary functioning.

The **Pleurectomy/Decortication**, or "P/D", is a less radical procedure because it involves the surgical removal of the tumor and the pleura, but not the lung. The procedure takes longer to perform as the surgeon carefully separates the tumor from the lung and repairs any damage to the diaphragm so that the patient will continue to enjoy the use of the

Because the only FDA approved chemotherapy for mesothelioma is approved only for patients who are not candidates for surgery, many oncologists believe it is prudent to refer the patient to a thoracic surgeon before starting chemotherapy. If the surgeon determines that surgery is an option, the surgeon and oncologist will work together to determine the appropriate role of chemotherapy in the patient's treatment.

III. Radiation

Radiation Therapy (or "radiotherapy") involves the localized use of high-dose radiation on pleural mesothelioma cancer cells. It works by destroying the cancer cells in the affected area.

The treatment is divided into several sessions, usually one session a day for five days with a break at the weekend. This ensures that less damage is done to healthy cells than to cancer cells. The damage to healthy cells is usually temporary, but is the reason that radiotherapy has some unwanted side effects such as fatigue and skin changes.

Radiation therapy can both reduce the size of a tumor and relieve symptoms like pain and shortness of breath. It is frequently used after surgery in order to kill the cancer cells which remain after the tumor is removed. It can also be combined with chemotherapy to combat the metastatic spread of the disease. However, doctors will limit its use depending on the volume of the tumor and how close it is to other vital organs.

IV. New Approaches to Mesothelioma Treatment

In addition to these conventional treatments, there are a number of experimental therapies which are still being tested for safety and effectiveness. Depending on their eligibility, patients may be able to try an experimental treatment by participating in a clinical trial. Several types of experimental therapy are briefly described here.

Angiogenesis Therapy cuts off the blood supply to tumors to slow their growth. **Gene Therapy** changes the genetic composition of living cells in order to fight cancer. **Immunotherapy** attempts to "trick" the body into recognizing that cancer cells are not normal by introducing drugs called biological response modifiers into the body. There are also new drugs in development that are targeted at the types of cells found in mesothelioma.



How to Choose a Treatment for Mesothelioma?

Patients who have recently been diagnosed with mesothelioma may feel overwhelmed and stressed. An insidious tumor has been growing in their body and time is of the essence in order to effectively treat it. There are many treatment options which may be available, but how does one navigate through the medical jargon and statistics in order to decide what to do?

The following is a four-step guide to help patients make informed decisions about their medical treatment under these very difficult circumstances.

Step 1: Find the Right Specialist

In order to quickly and effectively compare mesothelioma treatment options, the first thing a patient should do is find a doctor who specializes in the treatment of mesothelioma. Patients should ask their general practitioner, pulmonologist or oncologist to recommend a doctor who has established him or herself as an expert or specialist in treating mesothelioma. Specialists will typically have treated a hundred or more mesothelioma patients and have published articles on the subject of mesothelioma.

Mesothelioma specialists will have superior knowledge of the unique characteristics of mesothelioma, the latest and most innovative treatments and the advantages and disadvantages of available treatment options. They typically will also have greater knowledge about the tests which are necessary to quickly determine which treatments are appropriate for the patient.

The Pacific Heart, Lung & Blood Institute's website, www.phlbi.org is an excellent source of information on specialists, resources and treatment options for mesothelioma.

Step 2: Learn about Various Treatment Options

Once a patient has consulted with one or more specialists, it is likely that multiple treatment options will be presented, including those which the treating pulmonologist and oncologist may have proposed. It then becomes necessary for the patient to weigh the pros and cons of each alternative.

The most valuable resource will, again, be the specialist. However, because the specialist is typically very busy, it is important for the patient to make the most of every visit. One way to do this is to write down all of the alternatives that have been presented and go through the list with the

specialist, having the specialist provide his or her assessment of the pros and cons of each treatment in terms of risks, side effects, recovery times, survival, quality of life and cost. It is wise to have another family member or trusted friend in attendance to help write down the doctor's opinions.

Step 3: Talk with Others

Patients and families who have gone through the same decision-making process regarding alternative mesothelioma treatments are an invaluable resource. They can explain how they reached their decision, why they that multiple treatment options will be presented, including those which the treating pulmonologist and oncologist may have proposed. It then becomes necessary for the patient to weigh the pros and cons of each alternative.

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Step 3: Talk with Others

Patients and families who have gone through the same decision-making process regarding alternative mesothelioma treatments are an invaluable resource. They can explain how they reached their decision, why they chose one treatment plan or doctor over another and, most importantly, they can explain what might be expected in terms of side effects and recovery time.

The Pacific Heart, Lung & Blood Institute has helped many newly diagnosed patients make important treatment decisions by putting them in touch with other patients who have recently completed the treatments which are being considered. To speak with patients in your area, contact the Institute at (310) 478-4678 or info@phlbi.org.

Step 4: Find Out How Much it Will Cost

Finally, before embarking on any treatment plan, call your insurance company to determine which types of treatments are covered under your insurance plan as well as applicable deductibles which must be paid. Some insurance companies consider some of the more recent treatments for mesothelioma to be "experimental" and not subject to coverage. In addition, depending on the type of coverage, some insurers may not agree to cover treatments at University hospitals, where many specialists practice. If an insurer is denying coverage, the patient should find out what the anticipated cost will be to have the treatment performed outside of insurance. The family will then need to decide whether family resources or the anticipated recovery in an asbestos injury lawsuit will be sufficient to cover the cost.

One Specialist's Approach to Treating Mesothelioma:



"Remove the tumor, save the lung and treat the disease as a chronic illness."

- Dr. Robert Cameron

Dr. Robert Cameron is a cardiothoracic surgeon and surgical oncologist who specializes in the treatment of malignant pleural mesothelioma. He is the director of the mesothelioma program at the David Geffen School of Medicine at the University of California, Los Angeles (UCLA) and the chief of thoracic surgery at the West Los Angeles Veterans' Administration Medical Center.

Dr. Cameron has been treating mesothelioma patients for over 20 years. He is the innovator of the lung-sparing Pleurectomy/Decortication surgical procedure for mesothelioma and has performed the procedure on over 300 patients to date. He has also published many articles and is a frequent lecturer on the surgical and multi-modal management of pleural mesothelioma.

Treating Mesothelioma Like Other Chronic Diseases

"Despite all of the advances in modern medicine, there are a number of diseases or illnesses which doctors simply can't cure. Diabetes and high blood pressure are good examples. But through treatments which control these conditions, patients are able to live with them for extremely long periods of time."

Mesothelioma is another disease for which there is currently no 'cure'. No surgery, drug or other treatment can completely remove or destroy all of the mesothelioma cancer cells. Until a 'cure' can be found, it is our approach at UCLA to control mesothelioma and provide the patient with the best possible quality of life.

One Specialist's Approach to Treating Mesothelioma: Dr. Robert Cameron Continued

We attempt to do this through multi-modal treatment beginning with the lung-sparing Pleurectomy/Decortication ("P/D") surgery, followed by radiation and then a long-term maintenance therapy utilizing immunotherapy and a combination of other conventional and experimental treatments designed to stop the recurrence of the tumor.

Mesothelioma is Much Different Than Other Cancers

"Most other cancers grow as a nodule or a lump in an organ. This often allows a surgeon to remove the disease entirely by getting around the tumor with enough extra normal tissue to get what is called a margin.

Mesothelioma is much different. It is a sheet of cancer cells which virtually cover every organ and surface inside the chest. This prevents a surgeon from being able to remove the disease entirely. There simply isn't enough normal tissue surrounding the tumor to allow the surgeon to get a margin without interfering with major organs like the heart, lungs, major blood vessels, diaphragm and ribs.

Therefore, whether performing a P/D or Extrapleural Pneumonectomy ("EPP"), the goal of the surgery is to remove all visible tumor. Neither procedure is capable of removing all of the cancer cells."

P/D, not EPP, is the Preferred Surgical Treatment for Pleural Mesothelioma

"EPP is a very radical procedure in which a surgeon goes into the chest through a large incision and virtually cuts out the lung and strips the tumor off of all the surfaces. The lung is an innocent bystander underneath the tumor, but is removed along with the tumor in order to theoretically get better clearance.

P/D involves opening the chest and removing all visible tumor. Instead of removing the lung, you carefully separate the tumor from the healthy lung. This can be done very completely so it provides just as much tumor

One Specialist's Approach to Treating Mesothelioma: Dr. Robert Cameron Continued

clearance as taking out the lung in an EPP. It does so with much less sacrifice of lung function and risk of having surgical complications that result in bad outcomes and even death.

Another advantage of P/D over EPP is that P/D allows you to limit the operation site to the areas of tumor. An EPP often expands the surgery into new places where there is no tumor. This can cause the tumor to spread. In P/D, we leave barriers in the form of the diaphragm, pericardium and chest wall, between tumors and other areas to reduce the risk of spreading.

My philosophy has always been: 'do no harm.' Taking out a lung does harm and there is absolutely no benefit to the patient. Doing a bigger operation makes no sense from an oncology standpoint, because a surgeon cannot rid a mesothelioma patient of every last cancer cell and cure them – that won't happen. Once you accept that, then you can understand why P/D is the best surgical option with the least side effects, the least chance of dying and a better chance of getting as much tumor clearance as possible."

The Role of Post-Surgery Radiation and Maintenance Therapies

"Once the visible tumor has been removed, our attention turns to the cancer cells which inevitably remain at the surgery site. The patient undergoes a series of radiation treatments, each of which takes less than 15 minutes to complete. The goal of the radiation is to kill as many cancer cells as possible to prevent the reorganization of cancer cells and recurrence of a tumor.

The radiation oncologists at UCLA are very experienced in treating patients who have undergone a P/D where special care must be taken to focus the radiation on the sites of the cancer cells so that no damage is done to the healthy lung.

One Specialist's Approach to Treating Mesothelioma: Dr. Robert Cameron Continued

Following radiation, our patients begin long-term maintenance therapy with daily self-administered injections of an immunotherapy agent which is designed to stimulate the body's own immune system to prevent the remaining cells from developing into a tumor.

Our patients are then monitored closely with regular CT scans and follow-up visits every three to six months. At the first sign of tumor recurrence, we are able to utilize one of a number of treatments, such as radiation, chemotherapy and cryoablation to attack it. As more treatments are developed in the future, we will be able to add these to our arsenal of maintenance therapies."

Interested in Learning More?

If you have been diagnosed with malignant pleural mesothelioma and would like to learn more about Dr. Cameron's approach to treating the disease, or to find out if you are a candidate for the treatment, you may contact his office at **(310) 470-8980**.

*Dr. Cameron sees patients
in consultation at his
Los Angeles, California clinic located at:*

**10780 Santa Monica Boulevard
Suite 100
Los Angeles, California 90025-7613**

If you are traveling from out of town and would like assistance with travel arrangements, contact the Pacific Heart, Lung & Blood Institute at **(310) 478-4678** or info@phlbi.org.



Non-Medical Issues Confronting Mesothelioma Patients

Being diagnosed with mesothelioma raises a whole host of non-medical issues which the patient and family must address. Once the initial shock has passed, there will be questions and concerns about the practical aspects of handling the disease. The following are some of the most common concerns which people face after a mesothelioma diagnosis:

Will I Have to Quit My Job?

Working is an important issue for many patients. The decision to work is sometimes taken out of the patient's control depending on the severity of the disease and the treatment options which are selected. Even if the patient feels well enough to work, it should be realized that the rigors of work may deplete the energy which the body's immune system requires to continue fighting the disease. Ultimately, this will be an issue that is best addressed by the patient and his or her doctor.

Can I Be Fired Because of My Illness?

It is illegal for an employer to fire or lay off an employee because of an illness such as mesothelioma. If this happens, the patient should seek immediate legal help.

How Will My Illness Affect My Daily Life?

Every patient is different and it is difficult to determine how mesothelioma and the treatments which are received may affect the patient's daily life. Dealing with an illness like mesothelioma, which tasks the body's respiratory and immune system, often causes the patient to tire easily. Even the most routine tasks can seem overwhelming. It is important to rest, but at the same time, it is also important to engage in exercise in order to maintain endurance.

It is also important to eat. The body's immune system is working overtime and needs nourishment more than ever before. Unfortunately, the pressure exerted on the chest and abdomen as well as the effect of some

medications often cause a patient to lose his or her appetite. It is important for family members and caregivers to do everything they can, such as cooking the patient's favorite meals, in order to keep the patient eating.

Do I Have Any Legal Rights?

Exposure to asbestos is the only known cause of mesothelioma. Asbestos was the "miracle mineral" which was used in thousands of consumer, construction, industrial and automotive products for most of the 1900's. Because the hazards of asbestos were well known decades before manufacturers started removing it from their products, there are important legal rights which are possessed by most patients who are diagnosed with mesothelioma. The funds which are received from asserting these rights are often essential to dealing with the financial impact of mesothelioma.



Resources

The National Cancer Institute

[www.cancer.gov/cancertopics/
types/malignantmesothelioma](http://www.cancer.gov/cancertopics/types/malignantmesothelioma)

NCI's malignant mesothelioma section provides information on treatment published medical literature and clinical trials.

American Institute for Cancer Research

(800) 843-8114

www.aicr.org

Fosters research on diet and cancer prevention, interprets the evidence, and educates the public about the results.

American Cancer Society:

(800) ACS-2345

www.cancer.org

A nationwide, community-based voluntary health organization.

Cancer Hope Network

(800) 552-4366

www.CancerHopeNetwork.org

Provides free and confidential one-on-one support to cancer patients and their families.

Hospice Link

(800) 331-1620

www.hospiceworld.org

An organization that maintains a computerized database of all hospice and palliative care programs in the United States.

The Asbestos Disease Awareness Organization

www.asbestosdiseaseawareness.org

Founded by Linda Reinstein, wife of malignant mesothelioma patient Alan Reinstein.

The John McNamara Foundation

(818) 429-9431

www.thejohnmcnamarafoundation.org

Founded by TC McNamara, wife of malignant mesothelioma patient John McNamara.

LiveStrong

www.livestrong.org

Founded In 1997 Lance Armstrong, the foundation's mission is 'to inspire and empower' cancer sufferers and their families.

Air Charity Network

(877) 621-7177

www.aircharitynetwork.org

Provides access for people in need seeking free air transportation to specialized health care facilities or distant destinations due to family, community or national crisis.

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