



**Heart & Vascular  
Institute**

Westchester Medical Center Health Network

# An Introduction to the Westchester Medical Center **Pulmonary Hypertension Program**



Westchester Medical Center (WMC) in Valhalla, New York, and the WMCHHealth Heart and Vascular Institute, are proud to showcase the Pulmonary Hypertension Program, serving Westchester County and beyond. Its goal is to provide state-of-the-art advanced diagnosis and treatment for this rare condition. As a large tertiary center, WMC currently offers cutting-edge, personalized treatment for cardiovascular disease including advanced heart failure, valvular, coronary and vascular disease.



Julio A. Panza, MD  
Chief of Cardiology  
Westchester Medical  
Center and WMCHHealth

*“In continuing its mission to provide a high level of care to the region, WMCHHealth now introduces its multidisciplinary team dedicated to treating pulmonary hypertension. An overview of the components of the Pulmonary Hypertension Program is detailed in the following pages.”*

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**Director**  
**Gregg M. Lanier, MD**



**Cardiothoracic Surgery**  
**Ramin Malekan, MD**

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## The Pulmonary Hypertension Program Leadership

**Gregg M. Lanier, MD,** Director of Pulmonary Hypertension at Westchester Medical Center (WMC), is a heart failure specialist with training in pulmonary hypertension (PH). He completed his medicine residency and fellowship at Mount Sinai Hospital in New York City, and spent an additional year in advanced heart failure training at New York-Presbyterian Columbia Hospital. He has more than eight years of hands-on experience with the diagnosis and treatment of PH, including expertise with all drugs currently approved for this condition. He has published several articles on PH, and is a current principal investigator at WMC of a multicenter clinical trial in PH. Additionally, he participates in patient support groups, is a member of the Pulmonary Hypertension Association, and is active in the region promoting awareness of this disease.

**Ramin Malekan, MD,** is a cardiothoracic surgeon at WMC who trained previously at Mount Sinai Hospital in New York City. He sought additional training and experience in performing pulmonary thromboendarterectomy, a highly skilled surgical treatment of PH from chronic pulmonary blood clots. Working together, Dr. Lanier and Dr. Malekan, along with many other consultative services at WMC, are able to provide comprehensive and highly effective care for this condition.

## Overview of Pulmonary Hypertension

**Pulmonary Hypertension (PH)** is a general term for a multitude of conditions that result in elevated pressures in the lung vasculature. While common diseases such as heart dysfunction and emphysema may result in mildly elevated pressures incidentally found in routine testing, some patients develop very specific disease of the pulmonary arterioles, which causes pulmonary arterial hypertension (PAH). Untreated, this rarer form of PH can result in significant symptoms and a very poor prognosis.

The World Health Organization (WHO) has categorized the different causes of PH into five groups. PAH, or WHO group 1 PH, is an often misunderstood disease that can cause significant morbidity and mortality. Idiopathic PAH is rare, with an incidence of about 12 per 1 million people. This disease predominantly affects younger women, usually in their child-bearing years. Unfortunately, the diagnosis of PAH is made often more

than two years after the onset of symptoms, and this delay is thought to lead to overall poor outcomes. Within group 1 PAH, there are several conditions such as familial PAH, and PAH secondary to: liver disease (portopulmonary hypertension), scleroderma, HIV, diet drugs, congenital heart disease, methamphetamine use and schistosomiasis. More common causes of PH include non-PAH conditions such as WHO group 2 PH from left heart disease (heart failure), WHO group 3 PH from low oxygen levels, and WHO group 4 PH from chronic pulmonary emboli (chronic thromboembolic pulmonary hypertension or CTEPH). Other miscellaneous causes of PH are included in group 5, as shown in the table below.

The most important mission of a comprehensive PH program is to first establish the correct diagnosis, as the treatment depends on the cause. The diagnostic evaluation of a patient with PH requires multiple tests and specialists

working in consultation with a centralized PH program. After the diagnosis is established, the PH team determines the need and monitors the initiation of oral, inhaled, intravenous and subcutaneous medications as appropriate. In the case of WHO group 4 PH, surgical pulmonary thromboendarterectomy may result in a “cure” of the disease with excellent clinical outcomes.

This coordination of multiple disciplines in the diagnostic evaluation and treatment of patients with PH is the central purpose of an interdisciplinary PH program and is essential in ensuring quality outcomes such as improved quality and duration of life. The WMCHealth Heart and Vascular Institute’s Pulmonary Hypertension Program has a prominent and growing role in the highly specialized care of these patients who are referred from a large geographical and sometimes underserved region of New York, northern New Jersey, and western Connecticut.

### World Health Organization Classification (WHO) of Pulmonary Hypertension (PH)

Group 1	Group 2	Group 3	Group 4	Group 5
<b>Pulmonary Arterial Hypertension (PAH)</b> <ul style="list-style-type: none"> <li>• Idiopathic/Familial</li> <li>• Connective Tissue Disease (Ex. Scleroderma)</li> <li>• Portopulmonary Hypertension</li> <li>• HIV, Drugs and Toxins</li> <li>• Congenital Heart Disease</li> </ul>	<b>PH with Left Heart Disease</b> <ul style="list-style-type: none"> <li>• Systolic, Diastolic and Valvular Heart Failure</li> </ul>	<b>PH from hypoxia</b> <ul style="list-style-type: none"> <li>• COPD/emphysema</li> <li>• Asthma</li> <li>• Obstructive Sleep Apnea</li> <li>• Pulmonary Fibrosis</li> </ul>	<b>Chronic Thromboembolic PH (CTEPH)</b>	<b>Multifactorial</b> <ul style="list-style-type: none"> <li>• Hemolytic Anemias</li> <li>• Sarcoidosis</li> <li>• Sickle Cell</li> <li>• Systemic Disorders</li> </ul>

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## Diagnostic Evaluation of PH

Patients with possible PH are referred to the PH program at WMC at varying points in their evaluation and disease. A thorough assessment includes performing tests to screen for all possible risk factors for PH. Each of these tests requires coordination with several other specialties and departments. Not all of the suggested tests are indicated, and some may have been performed prior to referral. In the initial comprehensive consultation, prior test results are obtained and reviewed, and a plan for further testing is devised.

The initial consultation of a patient referred for PH usually starts in the outpatient cardiology office. The initial visit includes resting oximetry, ECG and a six-minute walk test. At subsequent visits, if familial PAH is suspected, genetic counseling and testing may be done to evaluate for mutations such as BMPR2 and ALK-1 PAH.

Several other specialists familiar with PH are essential in the initial evaluation and subsequent treatment. Referrals are often made to different departments at WMC, including: pulmonary (WHO group 3 PH from COPD, asthma, OSA, lung disease), infectious disease in PH-HIV, adult congenital heart disease, and hepatology in portopulmonary HTN (liver transplant may improve outcomes).

Heart failure specialists may be consulted in cases of WHO group 2 PH from systolic, diastolic or valvular heart failure. Clear lines of referral and communication are necessary for efficient evaluation and care in a comprehensive PH program.

Functional assessment with a cardiopulmonary exercise tolerance test is used to stratify risk for adverse outcomes and to distinguish between cardiac and pulmonary limitations of exercise.

This test, as well as pulmonary function tests, are performed in the Pulmonary Department and Laboratory at WMC. Sleep studies and overnight oximetry are indicated in patients with a history of snoring or hypoxia. This study may be done at WMC or sometimes in a patient's home. Clearly defining a possible WHO 3 aspect of PH is important before PAH medications are started, as inappropriate PAH therapy may worsen shortness of breath.





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## Testing

### Non-invasive Imaging and Testing

Perhaps the most important non-invasive test in PH is echocardiography. The non-invasive cardiology laboratory department, under the direction of Tanya Dutta, MD, has a dedicated PH protocol for estimating pulmonary artery pressure, quantifying right ventricular function, and using bubble contrast echocardiography when indicated to detect intra-cardiac shunts. Transesophageal echocardiography is sometimes employed in the evaluation of possible congenital heart disease.

Other cardiac imaging includes cardiac CT and cardiac MRI, both performed to elucidate congenital heart disease, including ASD, VSD, anomalous pulmonary veins, and other structural heart disease that could contribute to other etiologies of PH (Group 1 or 2 PH). The radiology department performs several different tests in the initial evaluation of PH. Nuclear imaging with a VQ scan is required to rule out CTEPH. CT of the chest, with or without contrast, is necessary to assess for acute pulmonary embolism, CTEPH and parenchymal lung disease. Ultrasound is sometimes used to look for blood clots in the legs and to test for liver disease.

### Invasive Testing

When the suspicion for PAH is very high, or when the non-invasive testing is completed and suggests WHO group 1 PAH, then the next essential test is a right heart catheterization (RHC). This is performed in the cardiac catheterization laboratory by invasive cardiologists who follow an established diagnostic protocol. This procedure may involve vasodilator testing with nitric oxide, volume loading in cases of suspected WHO 2 PH, and exercise testing. A thorough RHC is important to establish the correct diagnosis and understand the physiology prior to starting treatment.

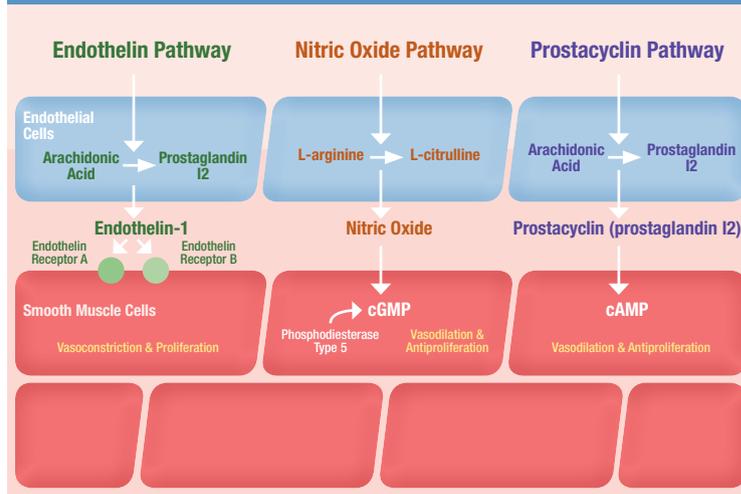
In cases of CTEPH, a pulmonary angiogram may be performed. This test is done by inserting a catheter into the pulmonary arteries and injecting IV contrast to take high resolution, real-time images of the blood vessels.

## PAH Drug Classification

Endothelin Pathway ETRA	Nitric Oxide Pathway PDE-5 Inhibitor / Guanylate Cyclase Stim	Prostacyclin Pathway
Bosentan	Sildenafil	Epoprostenol IV
Ambrisentan	Nitric Oxide (Inhaled) Acute Use	Treprostinil Sub-Q, IV, or Inhaled
Macitentan	Tadalafil	Iloprost Inhaled
	Riociguat	Selexipag (Oral IP Agonist)
		Orenitram (Oral Treprostinil)

ETRA = Endothelin receptor antagonists  
PDE-5 = Phosphodiesterase type 5

## Key Pathways Implicated in PAH Pathogenesis



## Treatment

### Outpatient Care

Initiation and continuation of treatment for WHO group 1 PAH is most often done in the outpatient setting. Treatment involves background therapy such as calcium channel blockers, supplemental oxygen, diuretics, digoxin and sometimes anticoagulation. There are currently 14 drugs approved for PAH treatment. The endothelin receptor antagonists include bosentan, ambrisentan and macitentan. The cyclic GMP/nitric oxide pathway drugs includes sildenafil, tadalafil and riociguat. Inhaled nitric oxide is only used in the inpatient setting. The third pathway, the prostacyclins, include epoprostenol (IV), thermostable epoprostenol (IV), treprostinil (oral, inhaled, subcutaneous and IV), iloprost (inhaled) and selexipag (oral IP agonist). Each of these drugs has side effects that require close monitoring and follow-up. Escalation and add-on therapy are recommended while a patient is monitored over time by echocardiograms, six-minute walk tests, blood tests and frequent clinical assessment.

Outpatient treatment with these PAH drugs requires 24-hour phone coverage by a PH specialist within the WMC PH Program. Mid-level practitioners are also essential in the day-to-day care of patients on PAH therapy, as several of the medications have side effects and diuretics need frequent adjustment. The WMC PH Program prides itself on ensuring timely accessibility to essential team members.

### Inpatient Care

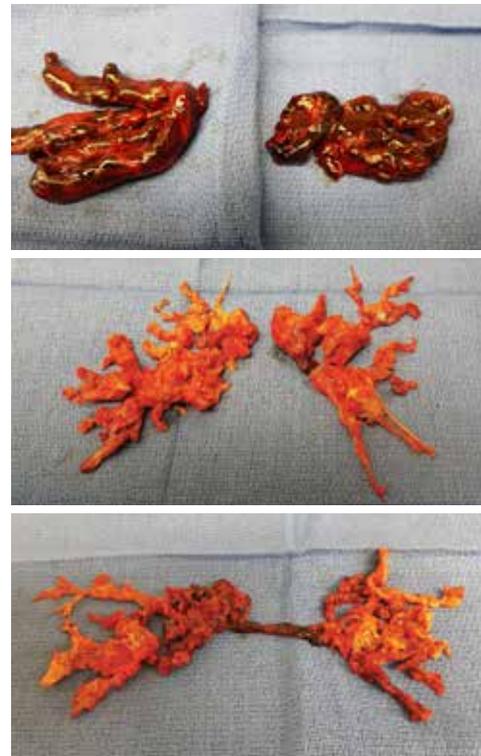
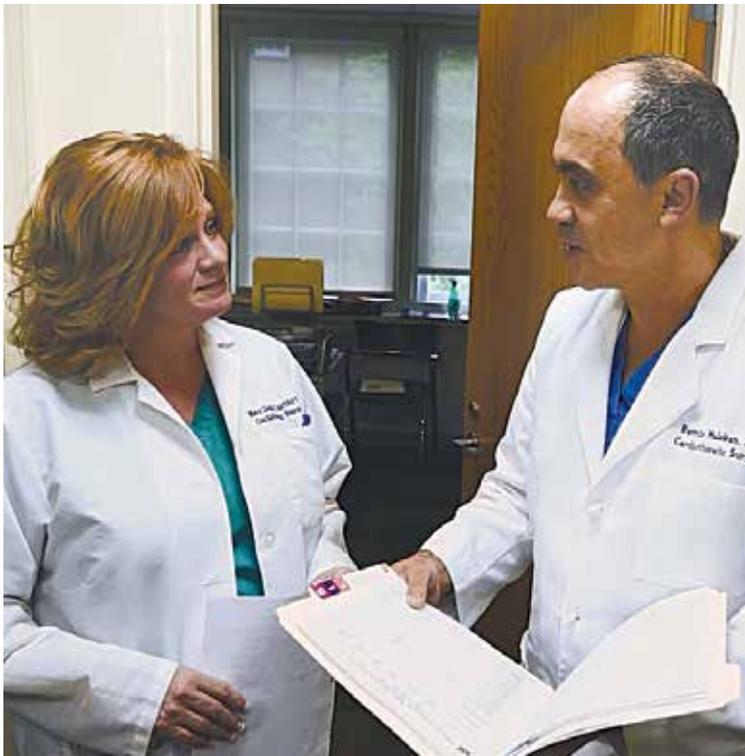
Our PH program has several outpatients on active PAH treatment, including IV or subcutaneous prostacyclins. If these patients become ill, they are admitted to WMC or a community hospital and require urgent transfer. At WMC, a team of nurses, physicians, pharmacists and other medical professionals have familiarity with these drugs and are able to safely start or restart IV or SQ prostacyclin therapy. The support of the pharmacy department, under the direction of Nadine Sarkissian, PharmD, is also central to the care of PH patients who are admitted. Timely preparation of these drugs is an important task of an experienced PH pharmacy department. Initiation of IV prostacyclin requires coordination with multiple different team members such as inpatient NPs, PH specialists, nurses and pharmacy. Placement of tunneled central venous access is performed by interventional radiology for long-term IV epoprostenol and treprostinil treatment. Nutrition, psychiatry, case management and social work are additional services that are often utilized during the inpatient setting.

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## Pulmonary Embolism and Chronic Thromboembolic Pulmonary Hypertension

CTEPH (WHO group 4 PH) is treated with pulmonary thromboendarterectomy (PTE) in surgical candidates, and by riociguat if the patient is not a surgical candidate. More than 20 PTEs have been completed at WMC with outstanding clinical outcomes. Ramin Malekan, MD, is an experienced cardiothoracic surgeon who has been essential in building the surgical division of the WMC PH Program. Many patients with unrecognized CTEPH present to the hospital with a new, acute pulmonary embolism (PE). Some of these cases demand emergency surgery to remove the blood clots. At the time of the

pulmonary thrombectomy, if the stigmata of CTEPH are found, the cardiothoracic surgeons at WMC have the training and expertise to convert the usually straightforward thrombectomy to a more complex and complete PTE. Very few centers in the United States have the ability to offer this extensive and complex surgical intervention for acute PE and CTEPH, and this aspect of the WMC PH Program makes it unique and attractive to the many hospitals in the area that care for massive and sub-massive PE patients that require transfer to a tertiary center.



Examples of specimens removed surgically by Dr. Malekan from the pulmonary arteries of three different patients. The image on the top is a large blood clot removed from a patient with massive PE. The specimen in the center photograph was removed from a patient with CTEPH who had systemic PH; the procedure resulted in normalization of pulmonary pressure. The image on the bottom is a specimen from a fresh blood clot associated with organized thrombi in a patient with CTEPH who presented with acute PE.



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## Extending the Impact of the Westchester Medical Center PH Program through Research

Large PH centers have a central role in the advancement of research into the treatment and understanding of this rare disease. Sachin Gupte, Md, PhD, and his team at New York Medical College, the Cardiovascular Translational Science Institute and National Institutes of Health are funding projects to study molecular markers, mechanisms and possible treatment modalities in PAH from scleroderma, based on blood specimens from research participants being treated at WMC. The researchers plan to develop a PAH animal model that reproduces human pathology by using blood samples from patients with scleroderma-PAH. Additionally, the team will attempt to isolate monoclonal antibody-producing immune and stem cells with the goal

to develop anti-idiotypic antibodies against Angiotensin II type-1 receptor and endothelin 1 type A receptor. This study will hopefully provide a bedside-to-bench approach to both the diagnosis and treatment of PH. WMC is also participating in a randomized, doubleblind, placebo controlled multicenter trial that will study the effect of an endothelin receptor antagonist, macitentan, in subjects with persistent PH after left-ventricular assist device placement for severe heart failure. Enrollment began at WMC in November 2017.

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## Team Members

### **Director**

Gregg M. Lanier, MD

### **Cardiothoracic Surgery**

Ramin Malekan, MD

### **Pulmonary Hypertension Providers**

Chhaya Aggarwal Gupta, MD  
Elliot (Avi) Levine, MD

### **Coordinators**

Kathleen McCrink, NP  
Carmela Musial, PA

### **Medical Assistant**

Victoria Curanaj, MA

### **Cardiac CT and MRI**

Anthon Fuisz, MD

### **Congenital Heart Disease**

Markus Erb, MD

### **Cardiac Catheterization**

Robert Timmermans, MD

### **Clinical Nurse Specialist**

Joan Kelly, RN

### **Echocardiography**

Tanya Dutta, MD  
Angelica Poniros, RCS

### **Heart Failure**

Alan Gass, MD

### **Hematology**

John Nelson, MD

### **Interventional Radiology**

Grigory Rosenblitz, MD

### **Laboratory**

John Fallon, MD

### **Liver Disease**

Roxana Bodin, MD

### **Nuclear Radiology**

Perry Gerard, MD

### **Palliative Care**

Michael Frankenthaler, MD

### **Pharmacy**

Nadine Sarkissian, PharmD

### **Pulmonary**

Dipak Chandy, MD

### **Research**

Sachin Gupte, MD, PhD  
Michael Wolin, PhD

### **Rheumatology**

Julia Ash, MD

### **Sleep Medicine**

George Maguire, MD



The WMCHealth Heart and Vascular Institute Pulmonary Hypertension Program involves a large team and several departments at Westchester Medical Center, uses interdisciplinary care coordination to establish the correct diagnosis, and then initiates specialized treatment in the outpatient and inpatient setting. Our program has the expertise to offer each patient with PH a very personalized and comprehensive evaluation and treatment that is unmatched in our region.

**For referrals for an outpatient evaluation, please call 914.909.6900 and fax relevant records to 914.493.2828, attention: Pulmonary Hypertension Program, WMC.**

**For urgent referrals, please email [Gregg.Lanier@WMCHealth.org](mailto:Gregg.Lanier@WMCHealth.org).**

**For requests for in-patient transfers or for emergencies, contact the transfer center for Westchester Medical Center at 914.493.5555.**



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