

Primary Pulmonary Hypertension Tutorial

Primary Pulmonary Hypertension is not a part of systemic hypertension but is presented here because of the close association of the name. To see this tutorial as it appears in our EMR, click [here](#)

Hypertension Evaluation

Primary Hypertension
Primary hypertension has no clearly defined etiology.

Secondary Hypertension
Most commonly hypertension has no symptoms!!!

Primary Pulmonary Hypertension

Signs & Symptoms

- Headache
- Dizziness
- Fatigue
- Palpitations
- Malaise
- Nosebleeds
- Weight Loss
- Swelling in the Legs
- Weakness
- Frequent Urination
- Increased Thirst
- Enlarged, Round, or "Moon" Face
- Excess Body and Facial Hair

Causes

Adrenal

- [Primary Aldosteronism](#)
- Cushing's Syndrome
- [Pheochromocytoma](#)
- Congenital Adrenal Hyperplasia

Endocrine

- Acromegaly
- Hypercalcemia
- Hyperparathyroidism
- Hyperthyroidism
- Hypothyroidism
- [Oral Contraceptives](#)
- Pregnancy-induced Hypertension

Neurologic

- Brain Tumor
- Bulbar Poliomyelitis
- Intracranial Hypertension

Drugs & Toxins

- Alcohol
- Cocaine
- Cyclosporin
- [Erythropoietin](#)
- Adrenergic Medications

Renal

- Renal Parenchymal Disease
- Polycystic Kidney Disease
- Urinary Tract Obstruction
- Renin-producing Tumor
- [Liddle Syndrome](#)
- [Renovascular Hypertension](#)

Vascular

- Coarctation of Aorta
- Vasculitis
- Collagen Vascular Disease

Information

- [Arterial Hypertension](#)
- [Renovascular Hypertension](#)

Return

- **Definition** -- Pulmonary hypertension is when the blood pressure in the arteries of the lungs is abnormally high. "Primary" means that there are no other diseases of the heart or lungs causing the high blood pressure.
- **Alternative Names** -- Pulmonary arterial hypertension; Sporadic primary pulmonary hypertension; Familial primary pulmonary hypertension
- **Causes And Risk** -- The cause of primary pulmonary hypertension is unknown. Some cases are caused by a genetic defect.
 1. The small arteries of the lung narrow throughout the lungs. Pulmonary hypertension is the result of greater resistance to blood flow. As a result of the increased workload caused by this resistance, the right side of the heart becomes enlarged.
 2. This disease is rare. It affects more women than men.
 3. Pulmonary hypertension is the narrowing of the pulmonary arterioles within the lung. The narrowing of the arteries creates resistance and an increased work load for the heart. The heart becomes enlarged from pumping blood against the resistance.
 4. The goal of treatment is control of the symptoms, although the disease usually develops into congestive heart failure.

- **Symptoms**
 1. Shortness of breath during activity
 2. Chest pain
 3. Weakness
 4. Fatigue
 5. Fainting
 6. Lightheadedness during exercise
 7. Dizziness
 8. Leg Swelling

- **Signs and Tests**
 1. A physical examination shows enlargement of the veins in the neck, normal lungs, a heart murmur, enlargement of the liver, and swelling due to fluid retention in the tissues.
 2. Tests
 - a. ECG
 - b. Chest X-Ray
 - c. Echocardiogram
 - d. Pulmonary Function Tests
 - e. Nuclear Lung Scan
 - f. CT Scan of the Chest
 - g. Cardiac Catheterization
 - h. Pulmonary Arteriogram

- **Call Your Doctor If...**
 1. Chest pain develops
 2. You develop shortness of breath when active
 3. Shortness of breath worsens
 4. You experience other symptoms

- **Treatment**
 1. There is no known cure. The goal of treatment is to control symptoms. Some people respond to treatment with oral medications, such as calcium channel blockers and diuretics. Others may need drugs like intravenous epoprostenol.
 2. Recently, a new oral drug that blocks abnormal constriction of blood vessels has become available and shows promise in treating pulmonary hypertension.
 3. As the disease progresses, oxygen may be necessary. If all else fails, suitable candidates may be helped by lung or heart-lung transplantation.
 4. Most patients with primary pulmonary hypertension are treated at centers which specialize in the care of these patients.

Primary Pulmonary Hypertension

Definition

Pulmonary hypertension is when the blood pressure in the arteries of the lungs is abnormally high. "Primary" means that there are no other diseases of the heart or lungs causing the high blood pressure.

Alternative Names

Pulmonary arterial hypertension; Sporadic primary pulmonary hypertension; Familial primary pulmonary hypertension

Causes And Risk

The cause of primary pulmonary hypertension is unknown. Some cases are caused by a genetic defect.

The small arteries of the lung narrow throughout the lungs. Pulmonary hypertension is the result of greater resistance to blood flow. As a result of the increased workload caused by this resistance, the right side of the heart becomes enlarged. Eventually, progressive heart failure may develop.

This disease is rare. It affects more women than men.

Pulmonary hypertension is the narrowing of the pulmonary arterioles within the lung. The narrowing of the arteries creates resistance and an increased work load for the heart. The heart becomes enlarged from pumping blood against the resistance.

The goal of treatment is control of the symptoms, although the disease usually develops into congestive heart failure.

Symptoms

- Shortness of breath during activity
- Chest pain
- Weakness
- Fatigue
- Fainting
- Lightheadedness during exercise
- Dizziness
- Leg Swelling

Signs and Tests

A physical examination shows enlargement of the veins in the neck, normal lungs, a heart murmur, enlargement of the liver, and swelling due to fluid retention in the tissues.

- ECG
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- Pulmonary Function Tests
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Call Your Doctor If...

- Chest pain develops
- You experience other symptoms
- You develop shortness of breath when active
- Shortness of breath worsens

Treatment

There is no known cure. The goal of treatment is to control symptoms. Some people respond to treatment with oral medications, such as calcium channel blockers and diuretics. Others may need drugs like intravenous epoprostenol.

Recently, a new oral drug that blocks abnormal constriction of blood vessels has become available and shows promise in treating pulmonary hypertension.

As the disease progresses, oxygen may be necessary. If all else fails, suitable candidates may be helped by lung or heart-lung transplantation.

Most patients with primary pulmonary hypertension are treated at centers which specialize in the care of these patients.

OK Cancel

Screening and Diagnostic Studies for Secondary Hypertension

Return

Suspicious

- Coarctation of Aorta
- Cushing's Syndrome
- Primary Aldosteronism
- Pheochromocytoma
- Renovascular Disease
- Thyroid Disease
- Hyperparathyroidism
- Renal Parenchymal Disease

Actions

- 24-hour Urinary Free Cortisol
- 24-hour Urine Protein and Creatinine Levels
- 2D Echo, Aorta
- Abdominal CT
- Aldosterone Excretion Rate During Salt Loading
- Adrenal CT
- Angiography
- Aortogram
- Calcium and Phosphorus Levels
- Captopril Renography
- Chest Film
- Clonidine Suppression Test
- Computed Tomography
- Dexamethasone Suppression Test
- Iodine 131 Metaiodobenzylguanidine Scan
- Iothalamate Glomerular Filtration Rate
- MRI, Aorta
- MRI, Renal Artery
- Plasma Aldosterone:Renin Ratio
- Plasma Catecholamines
- Renal Biopsy
- Renal Duplex Sonography
- Renal Ultrasound
- Renal Vein Renin Ratio
- Serum Calcitonin Level
- Serum Parathyroid Hormone Level
- Serum Thyroid Hormone Level
- Thyrotropin
- Urine Catecholamines
- Primary Pulmonary Hypertension
- Cardiac Catheterization
- Chest CT
- ECG
- Echocardiogram
- Nuclear Lung Scan
- Pulmonary Arteriogram
- Pulmonary Function Testing

If you think the patient has **Primary Pulmonary Hypertension**, check the box beside that name and the following will automatically have the boxes beside them checked:

- Chest Film
- Chest CT
- ECG
- Echocardiogram
- Nuclear Lung Scan
- Pulmonary Arteriogram
- Pulmonary Function Testing