



PULMONARY HYPERTENSION

by *Bill Wojciechowski, MS, RRT*

Pulmonary hypertension is an uncommon vascular condition in which the pulmonary vessels become narrowed, and the blood pressure in the pulmonary circulation increases. Normal pulmonary artery systolic pressure ranges from 20 to 30 mm Hg, the diastolic pressure varies from 6 to 15 mm Hg, and the mean pulmonary artery pressure is about 10 to 20 mm Hg. Compare these vascular pressures to those in the aorta of the systemic vasculature. The low vascular pressures stem from the vast cross-sectional area encompassed by the pulmonary circulation, the short distance traversed across this vascular network, the architecture of the right ventricle and the pulmonary vessels, and the physical mechanisms known as recruitment and distention.

PULMONARY VASCULAR ANATOMY

By virtue of their structure and architecture, the components of the pulmonary circulation help render this vascular network to be

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a low-pressure and a low-resistance circulatory system. For example, the walls of the right ventricle, which serves as the pump for the pulmonary circulation, are approximately one-third as thick as those of the left ventricle. The wall of the main pulmonary artery is roughly

one-fifth the thickness of the ascending aorta. Smooth muscle fibers constitute only about 2% of the thickness of the pulmonary artery wall. Interestingly, however, these muscle fibers constitute a much greater portion of the wall thickness for several ensuing generations of vessels. Consequently, arteries much smaller than the main pulmonary artery are relatively more muscular than the main pulmonary artery itself. The maximum relative amount of smooth muscle fibers emerges in arteries ranging from 100 μ m to 200 μ m in diameter. Intriguingly, these vessels reside in proximity to the respiratory bronchioles, where the conducting zone, or anatomic dead space, transitions into the gas exchange region.

As subsequent generations of these pulmonary vessels emerge, the smooth muscle fibers diminish as the small arteries evolve into pulmonary arterioles where ultimately the vascular smooth muscle fibers completely disappear. Consider the contrast between these arterioles and their counterparts in the systemic circulation. Systemic arterioles employ their amount of smooth muscle fibers to divert systemic arterial blood flow to organs or body systems experiencing the greatest metabolic demands.

As the pulmonary arterioles continue branching, they extend into single vessels 13 μ m in diameter. Each of these vessels furnishes blood to each of the lung's 300 million alveoli. These vessels ultimately give rise to about 100 billion pulmonary capillaries, each of which averages 12 μ m in length and 7 μ m in diameter.

PATHOGENESIS

Any condition that reduces the cross-sectional area of the pulmonary circulation, thereby limiting the recruitment and distention of pulmonary vessels, will produce pulmonary hypertension. Numerous factors contribute to the development of this vascular disease. They include (1) obstruction of pulmonary vessels (e.g., pulmonary emboli), (2) elevated left atrial pressure (mitral stenosis), (3) thickening of pulmonary arterial and arteriolar walls (scleroderma), (4) derangement and destruction of alveolar walls with damage to pulmonary capillaries in proximity (interstitial lung disease), (5) left-to-right shunting increasing blood flow in the pulmonary circulation (congenital cardiac anomalies), and (6) pulmonary vasoconstriction caused by hypoxemia, hypercapnia, and acidemia (COPD).

What all these conditions have in common is imposing an increased workload on the right ventricle subsequent to a reduction in the pulmonary vascular cross-sectional area. The reduced vascular cross-sectional area, whether caused by vessel thickening, obstruction, or destruction, leads to an elevation of pulmonary vascular pressures, increased right ventricular work, right ventricular dilatation or hypertrophy, and ultimately right heart failure.

PATHOLOGY

Despite pulmonary hypertension having numerous causes, the pathologic features are comparable. The primary pathologic presentation is thickening of pulmonary vessels resulting from muscularization of the walls of the pulmonary vessels. Normally, pulmonary arterioles are devoid of mural smooth muscle; however, chronic pulmonary vasoconstriction, as seen in COPD and cystic fibrosis, causes the arterial smooth muscle to extend into the arterioles. The smooth muscle in the medial layer of the pulmonary arteries undergoes hypertrophy, and the intimal lining of the arteries and arterioles experiences fibrosis. The net effect of this increased muscularization and fibrosis narrowing of the vascular lumen, producing increased resistance to flow, and increasing right ventricular afterload.

Somewhat of a pathologic variation to chronic pulmonary vasoconstriction is pulmonary venous hypertension, consequential to mitral stenosis or left ventricular failure. Chronic pulmonary venous hypertension leads to the acquisition of smooth muscle by

the pulmonary veins, which normally lack smooth muscle. Pulmonary veins evolve to resemble arteries. The pulmonary arteries themselves also respond to venous pulmonary hypertension by developing medial hypertrophy, which is present in patients with longstanding mitral stenosis.

In severe pulmonary hypertension, medial and intimal fibrosis often occurs in arteries. As severe pulmonary hypertension persists, necrosis of arterial walls may result. This development arises from intense contraction of smooth muscle within the medial layer which compromises oxygenation of the medial layer.

SIGNS AND SYMPTOMS

In the early stages of pulmonary hypertension, no specific signs or symptoms surface. Not until this vascular condition progresses, after months or even years, do signs and symptoms emerge. Generally, the first symptom to develop is shortness of breath. This presentation, i.e., dyspnea on exertion, usually manifests itself while the person performs activities of daily living such as taking out the garbage or climbing stairs. With the eventual progression of pulmonary hypertension, dyspnea at rest occurs. Other attending symptoms include dizziness, fainting (syncope), fatigue, and angina-like chest pain. Peripheral cyanosis, and swelling of the lower extremities (peripheral edema) and of the abdomen (ascites) appear as the afterload on the right ventricle increases.

TYPES OF PULMONARY HYPERTENSION

Two types of pulmonary hypertension can develop - primary hypertension and secondary hypertension. Primary hypertension, rare and less common than secondary hypertension, is inherited, and also develops from no known cause. Approximately, only 300 new cases of primary pulmonary hypertension are diagnosed per year. Because the signs and symptoms of primary pulmonary hypertension are nonspecific, diagnosis of this condition is difficult. Generally, many causes of secondary hypertension must be excluded before primary pulmonary hypertension is diagnosed. Consequently, the time from symptom onset to diagnosis is about 2 years.

Secondary pulmonary hypertension, on the other hand, arises from a variety of causes. For example, secondary pulmonary hypertension can be caused by chronic obstructive lung disease (chronic bronchitis and emphysema), left ventricular failure, mitral stenosis, obstructive sleep apnea, pulmonary embolism, scleroderma, neuromuscular disease, and interstitial lung disease. Diagnosing secondary hypertension involves diagnosing the underlying cause of the hypertension.

TREATMENT

The short-term (eight to 12 weeks) use of IV or injected prostacyclin has shown some benefits for treating pulmonary hypertension; however, evidence does not support its use on a long-term basis. Short-term benefits include increase exercise tolerance and improved blood flow. Other forms of treatment include inhalation of nitric oxide, and the use of calcium channel blockers (vasodilators). Oxygen therapy is often prescribed to improve the patient's oxygen status, and to possibly lessen the degree of associated hypoxemic vasoconstriction when present. Anticoagulants reduce the risk of blood clotting, and improve blood flow. Diuretics remove excess fluid which produces edema, and increases the work of breathing. Sildenafil has proved beneficial as it relaxes pulmonary vascular smooth muscle, and produces pulmonary vessel dilatation.

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Epoprostenol, which is similar to prostaglandin, is used to treat primary pulmonary hypertension when calcium channel blockers are ineffective. This drug removes blood lipids, lowers blood pressure, and prevents blood clots. Bostenan (Tracleer) is another prostacyclin-like medication. It, however, blocks the activity of the hormone endothelin, which is abundant in patients with primary pulmonary hypertension, and damages pulmonary vessels. Bostenan counters the effects of endothelin.

Lung transplantation is an accepted form of treatment for patients who fail medical therapy. It is the only treatment that completely resolves pulmonary hypertension. In fact, the right ventricle normalizes following lung transplantation. However, the five-year survival rate for lung transplant patients is approximately five years. Few patients are able to survive as long as 10 years.

Unfortunately, the prognosis for pulmonary hypertension is bleak. Most people succumb to this condition two to five years following diagnosis.

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