Approach to Internal Medicine

A Resource Book for Clinical Practice

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Pulmonary Hypertension

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WHO CLASSIFICATION OF PULMONARY HYPERTENSION

GROUP I. PULMONARY ARTERIAL HYPERTENSION

- **IDIOPATHIC**—primary
- **FAMILIAL DISORDERS**
- **DRUG AND TOXIN INDUCED**
- **PAH ASSOCIATED WITH SPECIFIC DISORDERS**—connective tissue disease, HIV, portal hypertension, congenital heart disease, schistosomiasis
- **PAH LONG TERM RESPONDERS TO CALCIUM CHANNEL BLOCKERS**
- **PAH WITH SIGNIFICANT VENOUS OR CAPILLARY INVOLVEMENT**—pulmonary veno-occlusive disease, pulmonary–capillary hemangiomatosis
- **PERSISTENT PULMONARY HYPERTENSION OF NEWBORN**

GROUP II. PULMONARY VENOUS HYPERTENSION DUE TO LEFT HEART DISEASE—heart disease with preserved LVEF, heart disease with reduced LVEF, valvular heart disease, cardiovascular conditions leading to postcapillary PH

GROUP III. PULMONARY HYPERTENSION DUE TO LUNG DISEASE AND/OR HYPOXEMIA—obstructive lung disease, restrictive lung disease, mixed restrictive/obstructive disease, including obstructive sleep apnea and obesity hypoventilation syndrome, developmental lung disease

GROUP IV. PULMONARY HYPERTENSION DUE TO PULMONARY ARTERY OBSTRUCTIONS—chronic thromboembolic, other pulmonary artery obstructions (i.e. tumor, parasites, foreign material)

GROUP V. PULMONARY HYPERTENSION WITH UNCLEAR AND/OR MULTIFACTORIAL MECHANISMS—hematological (pulmonary Langerhans cell histiocytosis, lymphangiomatosis), systemic and metabolic disorders (sarcoidosis), complex congenital heart disease, others (compression of pulmonary vessels by tumor, fibrosing mediastinitis)

PATHOPHYSIOLOGY

DEFINITION OF PULMONARY HYPERTENSION—mean pulmonary arterial pressure (PAP) >25 mmHg at rest or mean PAP >30 mmHg with exercise measured with right heart catheterization

CLINICAL FEATURES

HISTORY—unexplained dyspnea on exertion, cough, chest pain, hemoptysis, dizziness, syncope, hoarseness, past medical history (cardiac and respiratory diseases, thromboembolic diseases, HIV, cirrhosis, autoimmune and rheumatologic disorders), medications (amphetamine, diet pill such as dexfenfluramine)

PHYSICAL—vitals (tachypnea, tachycardia, atrial fibrillation, hypoxemia), peripheral cyanosis, small pulse volume, elevated JVP (prominent a wave or absent if atrial fibrillation, large v wave), right ventricular heave, loud or palpable P2, right-sided S4, tricuspid regurgitation murmur, Graham-Steell murmur (high-pitched, decrescendo diastolic rumble over LUSB), crackles, congestive liver, ascites, ankle edema

INVESTIGATIONS

BASIC

- **LABS**—CBC, lytes, urea, Cr, AST, ALT, ALP, bilirubin, INR, albumin, ANA, RF, anti-CCP, anti-Scl-70, anticentromere antibody, ESR, HIV serology, TSH
- **IMAGING**—CXR, CT chest, V/Q scan or CT chest PE protocol, echocardiogram

INVESTIGATIONS (CONT'D)

- **ECG**
- **OVERNIGHT POLYSOMNOGRAPHY**—if suspect OSA
- **ABG**
- **PFT**

SPECIAL

- **RIGHT HEART CATHETERIZATION WITH VASOREACTIVITY TESTING**

MANAGEMENT

SYMPTOM CONTROL—**diuretics**, **O_2**, **anticoagulation**, **calcium channel blockers** if positive vasoreactivity test (in high doses), **endothelin receptor antagonists** (bosentan, ambrisentan), **phosphodiesterase type-5 inhibitors** (sildenafil), prostacyclin analogues (epoprostenol, iloprost, selexipag), soluble guanylate cyclase stimulators (riociguat)

TREAT UNDERLYING CAUSE

ATRIAL SEPTOSTOMY

LUNG TRANSPLANT

REFERRAL TO A SPECIALIZED PULMONARY HYPERTENSION CLINIC

SPECIFIC ENTITIES

EISENMENGER SYNDROME—left-to-right shunt leading to pulmonary hypertension and eventually right-to-left shunt