

38. Pulmonary Hypertension

DEFINITION: Mean pulmonary artery ≥ 20 -25 mmHg rest or ≥ 30 mmHg with exercise.

Systolic PAP = 30-45 (mild), 45-60 (moderate), 60-70 (moderately severe), >70 mm (severe)

WHO-1973 Classification: 1. Arterial Plexiform, 2. Veno-occlusive, 3. Thromboembolic

PRIMARY:

1. Proliferating intimal hyperplasia of endothelial and smooth muscle proliferation of small pulmonary arteries and arterioles. Fibrosis and "Onionskin". Endothelial cell proliferation in the plexiform lesions and of hyperplastic smooth muscle cells in small pulmonary arteries with medial hypertrophy.
2. Similar findings with Pulmonary Hypertension due to Collagen Vascular Disease.
3. More common in females.

SECONDARY: (HYPOXIA begets Pulmonary Hypertension)

1. Cardiac disease-LV failure, Valvular (MR, MS, AI or AS) & Congenital (L->R shunts)
2. Pulmonary disease-Interstitial, obstructive, alveolar hypoventilation and s/p surgical pneumonectomy. (neoplasm, infection & sarcoid). Sleep apnea.
3. Arterial Occlusive- chronic thromboembolic pulmonary hypertension (CTEPH), IVDA, Parasitic.
4. Pulmonary veno-occlusive disease (PVOD) mediastinal adenopathy. pulmonary vein obstruction,
5. Miscellaneous-Portal HTN, HIV, hereditary, high altitude, hemoglobinopathies chemotherapy, (Rape Seed Oil, Fen-Phen)

1. HISTORY/SYMPTOMS:

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|---------------------|-------------|------------------|
| -CHF | -Dyspnea | -Lightheadedness |
| -Syncope/presyncope | -CP | -Cough |
| -Hemoptysis | -Hoarseness | -Fatigue |

2. PHYSICAL EXAMINATION:

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|------------------------------|-------------------|------------------------------------|
| -RV heave (Left parasternal) | -Right sided S4 | -Low volume carotid artery pulse |
| -Closely split S2 | -Loud S2 | -Large JVP <i>a</i> - wave |
| -Narrowed pulse pressure | -Tachycardia | -Large JVP v-wave (with severe TR) |
| -Right sided S3 | -TR or PI murmurs | -Cyanosis (due to L->R shunting) |
| -Edema | -Clubbing | -Ascites/hepatosplenomegally |

3. LABORATORY FOR PRIMARY PULMONARY HYPERTENSION:

- | | | |
|------|-------------------|---|
| -CT | -Open lung biopsy | --PFT's (Normal or restrictive with decreased DLCO) |
| -CXR | -Echocardiography | - Polysomnography |
| -ECG | -ABG | - Pulmonary angiography/CT |

4. BLOODS:

- CBC, LFT's, TSH, ESR, HIV, RF, ANA, ANA profile, SCL-70, ANCA, PT/PTT Antiphospholipid AB's
Consider obtaining a Protein C & S & Antithrombin III, Anti-Ku autoantibodies

FINDINGS:

ECG :-RVH, RAD, RBBB or IRBBB, P-pulmonale, S1Q3 or S1S2S3 ,normal QRS voltage and inferior strain.

CXR :-RV & RA enlargement, Main PA enlargement with tapering/pruning of the peripheral branches

CT:-Hypoperfusion parenchymal pattern, interstitial lung dz, thromboembolic dz, fibrosing mediastinitis.

ECHOCARDIOGRAPHY:-RVE, RAE, TR with high jet velocity, PI, D-shaped septum, small LV cavity size.

CARDIAC CATHETERIZATION (R & L): -Right Pressures, PVR, low CO. R/O shunts, test vasodilation.

TREATMENT:

1. Anticoagulation
2. O₂
3. Low dose Diuretics
4. Bosentan/TRACLEAR 62.5 to 125 mg bid, \$3000 per month)
5. PDE-5 Sildenafil/VIAGRA-REVATIO 20 mg tid
6. Flolan/Epoprostenol (Prostacylin-PGI₂) start 2 ng/kg/min Central Venous-3 min fatal withdrawal, \$58,000 per year.
7. Phlebotomy, Lung transplant
8. Others: Inhaled NO, Adenosine,
9. Nifedipine-xl 30 to 240 mg qd & Diltiazem.