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Pulmonary Hypertension

Definition: PA systolic ≥35 or PA mean ≥25 or PA diastolic ≥15

Symptoms: **Dyspnea on exertion**, chest pain, syncope

Etiology:

1. Pulmonary venous hypertension – most common form, usually due to left-sided heart disease
2. Chronic hypoxia (secondary vasoconstriction of vasculature) – common causes include:
    a. Long-term high altitude exposure
    b. Restrictive lung disease (obesity, pleural fibrosis, neuromuscular)
    c. Chronic upper airway obstruction (Obstructive sleep apnea)
    d. Chronic lower airway obstruction (chronic bronchitis, bronchiectasis, emphysema)
    e. Chronic diffuse parenchymal disease (insterstitial fibrosis, connective tissue disorders, rheumatoid, scleroderma)
3. Pulmonary artery obstruction (chronic thromboemboli, lupus, sickle cell)
4. Left to right shunts with increased flow across pulmonary vasculature (Extracardiac – patent ductus, intracardiac – VSD, ASD); Eisenmenger complex is reversal of shunt such that it flows right to left.
5. Idiopathic (Primary) – Female:Male (2:1), AAF:WF (4:1); mean age at diagnosis = 35 years; familial incidence of 6%
    a. Probable final common pathway from multiple etiologies
    b. Subtypes of arteriopathy (plexogenic, thrombotic), veno-occlusive, capillary hemangiomatosis
    c. Dexfenfluramine used for 6 months associated with 23x risk for development of pulmonary htn (absolute risk from exposure = 1:20,000)
    d. Associated with connective tissue diseases, especially CREST syndrome, lupus. Also HIV.

History:

a. Symptoms (DOE, chest pain, syncope), functional assessment (NYHA)
b. Search for secondary causes
    i. Connective tissue disease – **scleroderma/**CREST, lupus, rheumatoid
    ii. HIV
    iii. Meds – dexfenfluramine, fenfluramine, phentermine
    iv. Pulmonary embolism, sickle cell, congenital heart disease, elevated Left heart pressures, chronic hypoxemia

Physical Exam:

1. Loud pulmonic component of S_2 (audible at apex = loud)
2. Early systolic click (interruption of pulmonary valve opening into high pressure)
3. Midsystolic ejection murmur (turbulent flow across pulmonary valve)
4. Left parasternal lift (high RV pressure and RV hypertrophy)
5. Increased jugular “a” waves (high RV filling pressure)

Diagnostics:

1. Chest Xray – prominent pulmonary trunk and hilar pulmonary arteries; "pruning" or peripheral pulmonary arteries and obliteration of the retrosternal clear space
2. Echocardiography – Able to estimate PA systolic pressure indirectly; can measure RV size and evaluate for shunts.
3. Radionuclide scans – V/Q study useful to exclude chronic emboli
4. Pulmonary Arteriography – definitive 'gold standard' for emboli **BUT** may underestimate embolic burden if organized into vessel walls
5. Hemodynamics (Swan) – directly measures PA pressures, Cardiac output, PCWP

Prognosis:

1. Untreated survival – 1year 72%; 2 years 55%, 5 years 30%
2. Causes of death – RV failure (63%), pneumonia, sudden death
3. Eisenmenger complex live longer (up to 1-2 decades) with equivalent degrees of pulmonary htn

Treatment:

1. Search for secondary pulmonary hypertension and treat where applicable
    a. Pulmonary venous hypertension – mitral valve surgery (if stenotic)
    b. COPD – oxygen, ?lung reduction
    c. OSA – CPAP, weight loss
    d. High altitude – move to lower altitude
    e. Chronic thromboemboli treatment of choice = surgical **thrombectomy**
    f. Intracardiac shunt – heart/lung transplant or lung transplant with repair
    g. Vasodilators are contraindicated in pulmonary venous hypertension (cause pulmonary edema)
2. For Primary pulmonary hypertension, check for 'Responder' vs. 'Nonresponder' to vasodilator therapy (nonresponders have worse prognosis)
    a. Invasive hemodynamics with application of vasodilator (epoprostenol, adenosine, nitric oxide, calcium channel blockers)
    b. 25% are responders
    c. Initiate vasodilator therapy for responders
        i. Medications with studied symptomatic and survival data
            1. Nifedipine, verapamil, or diltiazem
            2. Epoprostenol (Flolan) – requires 24h IV administration (pump)
        ii. Invesitgational Medications
            1. Treprostenil – subcutaneous prostacyclin analogue
            2. Bosenten – FDA approved oral endothelin receptor antagonist
            3. Sitaxsenten – oral? Endothelin A receptor antagonist
            4. Sildenafil – oral Phosphodiesterase 5 inhibitor
3. All patients with primary pulmonary hypertension should receive coumadin unless otherwise contraindicated

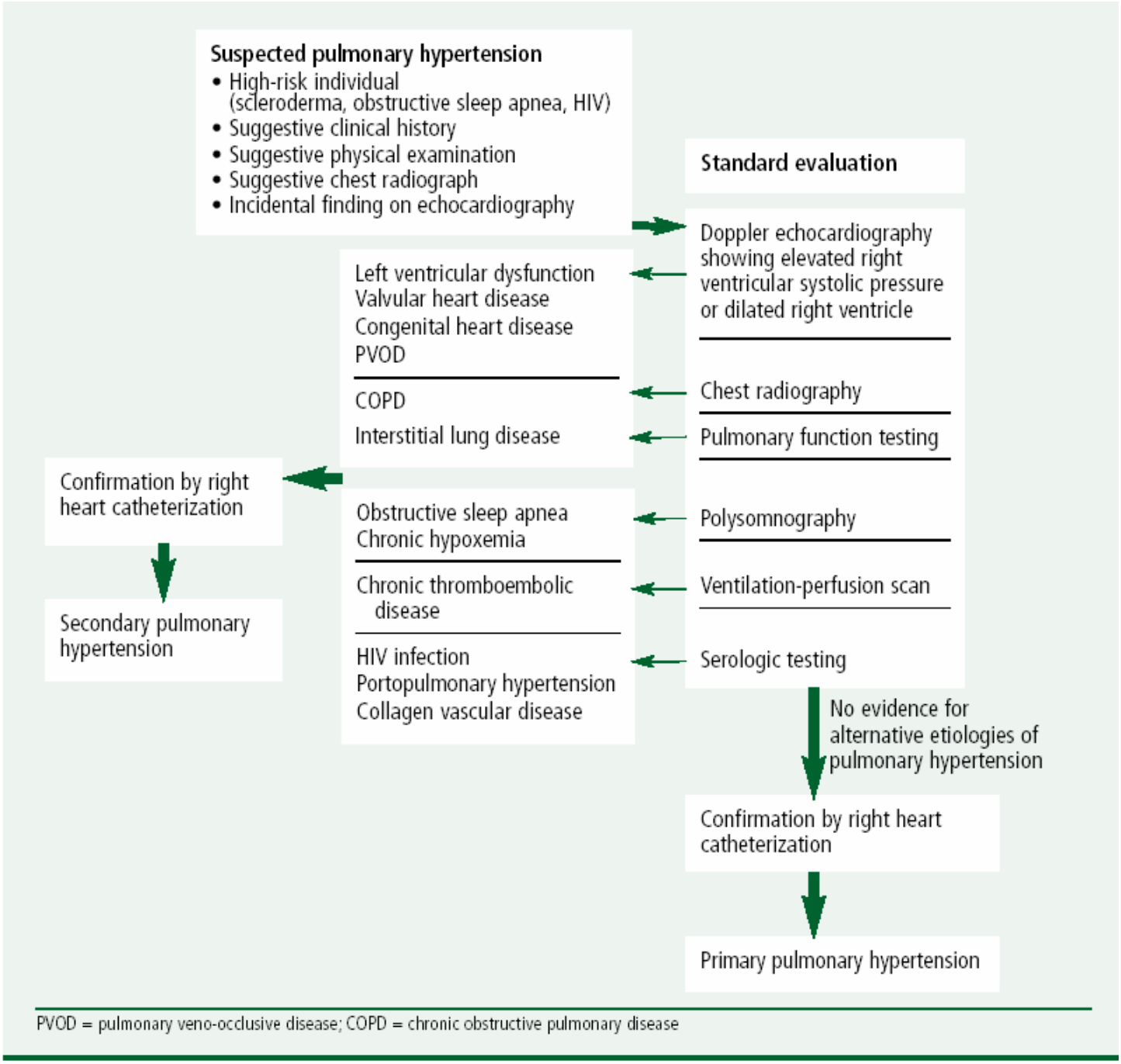

Figure 1. Clinical algorithm for the diagnosis of pulmonary hypertension.

Cleveland Clinic Journal of Medicine Supplement April, 2003: Diagnosis and Evaluation of Pulmonary Hypertension: http://www.ccjm.org/pdffiles/BUDEV.PDF

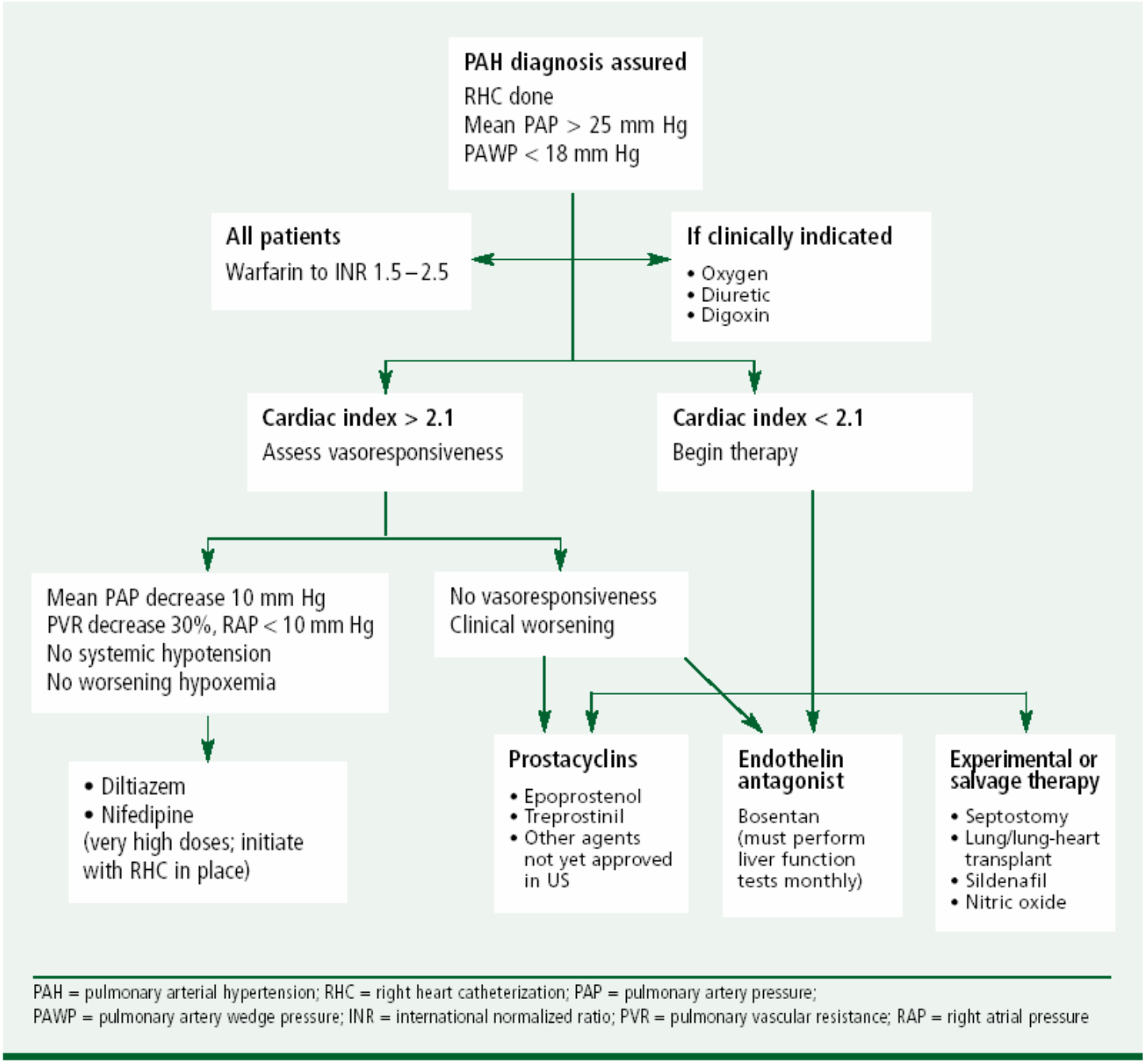

Figure 1. Algorithm for the management of patients with pulmonary arterial hypertension.

Cleveland Clinic Journal of Medicine Supplement April, 2003: Diagnosis and Evaluation of Pulmonary Hypertension: Treatments and strategies to optimize the comprehensive management of patients with pulmonary arterial hypertension: http://www.ccjm.org/pdffiles/GILDEA.PDF

References:

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