Cor Pulmonale

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Definition

- Alteration of the right ventricular structure or function that is due to pulmonary hypertension (PHTn) caused by diseases affecting the lung or its vasculature, airways, thorax, or respiratory control mechanisms.
- Excludes
  - Left sided heart disease
  - Congenital heart disease

Cor pulmonale may be acute or chronic.

The most common cause of acute cor pulmonale
Massive or multiple pulmonary emboli

Etiology of Cor Pulmonale (I)

- Lung and Airways
  - COPD
  - Asthma
  - Bronchiectasis
  - DILD (Diffuse Infiltrative Lung Disease)
  - Pulmonary tuberculosis
- Vascular Occlusion
  - Multiple Emboli
  - Schistosomiasis
  - Filariasis
  - Sickle Cell
  - P. Pulmonary Hypertension

Etiology of Cor Pulmonale (II)

- Thoracic Cage
  - Kyphosis > 100°
  - Scoliosis > 120°
  - Thoracoplasty
  - Pleural fibrosis
- N-M Disease
  - Polio Myelitis
  - Myasthenia Gravis
  - ALS (Amyotrophic lateral sclerosis)
  - Muscular Dystrophy

Etiology of Cor Pulmonale (III)

- Abnormal Respiratory Control
  - Idiopathic hypoventilation Syndrome
  - Obesity hypoventilation syndrome (Pick-Wickian syndrome)
  - Cerebrovascular disease

The most common cause of chronic cor pulmonale - COPD
**Pathophysiology**

Pathophysiological mechanisms causing pulmonary hypertension include:

- Pulmonary Artery vasoconstriction
  - Alveolar hypoxia
  - Blood acidosis
- Anatomic reduction of pulmonary vascular bed secondary to lung disorders
  - Emphysema
  - Pulmonary emboli
  - Increased blood viscosity
  - Erythrocytosis (includes polycythemia)
  - Sickle-cell disease
- Idiopathic primary pulmonary hypertension

**Pathophysiology**

Failure of right ventricle:

- **Pulmonary hypertension**
- **Myocardial anoxia**
- **Repeatedly pulmonary infection:** effect of bacterial toxin to the heart
- **Acid base disorder:** arrhythmia

**Pathologic Features**

- Lung: consistent with specific diseases
  - **Common Features:** hypertrophy of microvasculatures
- **Hallmark:** Right Ventricular Hypertrophy
  - 60g–200g, > 0.5 CM, RV/LV <2.5
  - Left Ventricular Hypertrophy
  - Hypertrophy of Carotid Body

**Patients with COPD**

- Most frequent cause of cor pulmonale
- Right ventricular hypertrophy (RVH) in
  - 49% of patients with FEV1 < 1.0 L
  - 70% of patients with FEV1 < 0.6 L
- Independent predictors of RVH
  - Hypoxemia
  - Hypercapnea
  - Erythrocytosis (not Polycythemia)

**Natural History**

- Several months to years to develop
- All ages from child to old people
- Repeated infections aggravate RV strain into RV failure
- Initially responds well to therapy but progressively becomes refractory
Symptoms of CP
- Directly attributable to PHTn
  - Dyspnea on exertion, fatigue, lethargy
  - Chest pain, syncope with exertion
- Typical exertional angina
  - Occurs in patients with primary or secondary PHTn even in the absence of epicardial CAD
  - Subendocardial RV ischemia induced by hypoxemia and increased transmural wall tension
  - Dynamic compression of left main coronary by enlarged PA
- Less common
  - Cough, hemoptysis, hoarseness
- With severe right ventricular (RV) failure
  - Anorexia, right upper quadrant discomfort

Physical Findings
- Cardiac findings
  - RV failure leads to systemic venous HTn
  - Elevated jugular venous pressure with a prominent V wave
  - RV S3
  - High pitched tricuspid regurgitant (TR) murmur
- Extra cardiac changes
  - Hepatomegaly, pulsatile liver
  - Peripheral edema—often related to hypercarbia and passive Na+ and water retention

Other Areas of Fluid Retention
- Pleural effusion, often bilateral
  - Right heart failure until proved otherwise
  - Also kidney and liver
- Engorged inferior vena cava
- Hepatic congestion
- Ascites
- Anasarca

Right Atrial Pressure Tracing

Jugular Pulsations
- A wave
  - RAP transmitted to jugular veins (JV) during right atrial systole
- V wave
  - Rise in RA and JVP due to continued inflow of blood to the venous system during late ventricular systole when the tricuspid valve is still closed
  - May also be elevated in heart failure and renal failure, but not cirrhosis.

Hepatojugular Reflux
- Assessed by applying firm sustained pressure over the upper abdomen with pt. breathing quietly.
- Response
  - Transient elevation by approximately 1 cm in normal response
  - In RHF sustained elevation
  - Low specificity and sensitivity
### Peripheral Edema

- **Edema formation requires**
  - Alteration in capillary hemodynamics that favors the movement of fluid from the vascular space into the interstitium (IS)
  - The retention of dietary or IV administered sodium and water by the kidneys.
  - Requires 2.5 to 3.0 liters of extra volume

- **Sequence of events**
  - Movement of fluid from vascular space into the IS reduces the plasma volume and consequently tissue perfusion
  - The kidney then compensates by retaining sodium and water

### Symptoms & Signs - Acute cor pulmonale

- **Sudden onset of severe dyspnea and cardiovascular collapse**
- Occurs in the setting of massive pulmonary embolism
  - Pallor
  - Sweating
  - Hypotension
  - Rapid pulse of small amplitude
  - Neck vein distention
  - Pulsatile distended, tender liver
  - Systolic murmur of tricuspid regurgitation along the left sternal border
  - Presystolic (S1) gallop

### Evaluation

- Laboratory CBC, chem.test, LFT’s, BNP
- Chest radiograph
- Electrocardiogram
- Two D and Doppler echocardiography
- Pulmonary function tests
- Radionuclide ventriculography
- Magnetic resonance imaging
- Right heart catheterization
- Lung biopsy

### Laboratory

- CBC-depressed or elevated Hgb, Hct
- Chem test-relationship of BUN to Creatinine
  - Normal ratio BUN/Cr approximates 20/1
  - Prerenal azotemia > 20/1
  - Intrinsic renal disease < 20/1
  - Estimated glomerular filtration rate (eGFR)
- Liver function tests
  - SGOT
  - SGPT

### Brain Natriuretic Peptide (BNP)

- A hormone released from myocardial cells
- Both atria and both ventricles
- Inhibits weakly
  - Renin-angiotensin system (Angiotensin II)
  - Endothelin secretion
  - Systemic and renal sympathetic activity
  - Plasma aldosterone production
BNP Continued

- Higher in
  - Older > younger
  - Women > men
  - Normal weight > obese
  - Renal failure
  - Congestive heart failure (right and/or left)
- Patient is his own reference point
  - Baseline
  - Post treatment

BNP Continued, Prognosis

- HF pts.- Highest quartile at baseline had higher mortality over 2 years at baseline (32.4 vs 9.7%) than lowest quartile.
- Following optimal medical treatment mortality increased proportionately to the level of the BNP elevation.

Normal Chest Radiograph

Radiograph in Cor Pulmonale

Radiograph and Cor Pulmonale

- Enlargement of Central PA’s
- In 95% of Pts with PHTn from COPD the diameter of the descending branch of the right PA is > 20 mm in width
- Peripheral vessels are attenuated leading to peripheral oligemia

Radiograph and Cor Pulmonale

Posteroanterior chest radiograph showing severe pectus excavatum and the complete displacement of the heart into left the hemithorax.

Lateral chest radiograph showing severe pectus excavatum and the complete displacement of the heart into the left hemithorax.
Normal Electrocardiogram

Right Atrial Enlargement on ECG

ECG in Cor Pulmonale

Two Dimensional Echocardiogram

Two D Echo, continued
Tricuspid Regurgitation (TR)
Doppler Echocardiography

- Most reliable noninvasive estimate of the Pulmonary Artery Pressure (PAP)
- Dependent on identifying an adequate tricuspid regurgitant jet
- More sensitive as PAP increases

2D Echo with Color Flow Doppler

Differentiating features between RHF with or without cor pulmonale/ pulmonary arterial hypertension

RHF without pulmonary hypertension
- Chest x-ray: Enlargement of pulmonary arteries (uncommon), oligemic peripheral lung fields (rare)
- Echocardiography: No evidence of increased pulmonary pressure. Septal flattening during diastole but not systole

Cor pulmonale/pulmonary hypertension present
- Chest x-ray: Right-sided cardiac enlargement, enlargement of pulmonary arteries, oligemic peripheral lung fields
- Echocardiography: Evidence of increased pulmonary pressure. Septal flattening during systole
- Physical examination: Evidence of underlying pulmonary pathology if cor pulmonale present (but not in primary PAH)

Pulmonary Function Testing (PFT’s)

- Primer and overview
- Satisfactory effort
- Obstruction
- Restriction
- Malingering

PFT Expiratory Maneuver

Expiratory Flow/Volume Loop
Right Sided Cardiac Catheterization

- When echo does not permit measurement of TR
- When symptoms are exertional and left sided pressures are unremarkable
- When therapy will be determined by precise measurement of pulmonary vascular resistance (PVR) and the response to vasodilators
- When left heart catheterization is also required (patients > 40 y/o and or with CAD)

Lung Biopsy

- Rarely, if ever required
- High risk procedure (elevated PVP, PAP)
- Transbronchial lung biopsy first
- Fiber optic thoracoscopy
- Never open thoracotomy

Acute cor pulmonale

- Arterial blood gas
  - Reduced partial pressure of arterial oxygen (PaO₂) due to ventilation-perfusion mismatch
  - Low partial pressure of carbon dioxide (PaCO₂) due to hyperventilation
- BNP and N-terminal BNP levels are:
  - Dramatically elevated in acute pulmonary embolism
- Spiral CT of the chest
  - Useful in diagnosing acute thromboembolic disease

Differential Diagnosis

- Coronary artery disease: Angina pectoris, Myocardial infarction history & Left ventricular hypertrophy (EKG)
- Rheumatic heart disease: History & echocardiogram
- Myocardial disease:
  - Without chronic respiratory disease
  - Echocardiogram
  - The total heart enlarged

Differential Diagnosis

- Liver cirrhosis
- Nephrotic syndrome
- Renal failure with significant volume overload

Complication:

- Pulmonary encephalopathy
- Acid-base disorder and electrolyte disturbances
- Shock
- DIC
- Arrhythmia
- Gastrointestinal hemorrhage
Prognosis of Cor Pulmonale

- When due to COPD, PHTn plus peripheral edema
  - 5 year survival 30%, mean 3 years from dx
  - Pulmonary vascular resistance >550 dynes/sec/cm rarely survive more than 3 years
  - May just reflect the degree of underlying COPD

Treatment

- **Oxygen**
  - Relieves pulmonary vasoconstriction
  - Decreases PVR
  - Increases RV stroke volume and cardiac output
  - Renal vasoconstriction may be relieved with increase in urinary sodium excretion
  - Improves arterial oxygen tension with enhanced delivery to
    - Heart
    - Brain
    - Other vital organs (kidneys)

Treatment-Diuretics

- Increasing RV filling volume using diuretics
  - Improve function of both RV and LV
  - RV dilatation is reduced LV filling improves
  - May improve cardiovascular performance
- Monitor for excessive volume depletion
  - BUN (blood urea nitrogen) "Prerenal"
  - Creatinine "Renal"
  - Estimated glomerular filtration rate (eGFR)
- Watch for metabolic alkalosis
  - May suppress ventilation

Complimentary Treatments Related to Severity of PHTn and its Systemic Effects

- Furosemide/Bumetanide/Torsemide
  - Loop diuretics
- Hydrochlorothiazide
  - Blocks sodium reabsorption
- Spironolactone/Eplerinone
  - Blocks aldosterone effect on both kidney and heart
- Angiotensin Converting Enzyme (ACE) inhibitor/ACE Receptor Blockers
  - Blocks Renin and Angiotensin
- Beta blockers (metoprolol, Atenolol, Carvedilol)
  - Blocks effect of norepinephrine

Treatment, Continued

- **Digoxin** is **NOT** indicated in pure CP
- Digitalis is used by some clinicians to support the failing right ventricle.

  **The indications of digitalis are:**
  1. uncontrolled heart failure therapy of diuretics after infection controlling and respiratory function improved;
  2. evident signs of right heart failure without severe infection;
  3. accompanied acute left heart failure.
Treatment, Continued

- These PA Vasodilators are of NO benefit
  - Hydralazine
  - Nitrates
  - Nifedipine
  - Verapamil

Theophylline/Terbutaline

- Has effects other than direct bronchial dilatation and diuresis
- Improves myocardial contractility
- Provides some degree of pulmonary vasodilatation
- Enhances diaphragmatic endurance
- Narrow range of efficacy

Phlebotomy

- When hematocrit > 55
- Goal is hematocrit < 50
- Secondary Erythrocytosis vs Polycythemia
- Treat underlying condition

Treatment

Acute exacerbation period

Controlling infection, clearing airways, elevating respiratory function, improving hypoxia and hypercapnia, and correcting respiratory failure and cardiac failure are the priority.

**Antibiotics:** commonly used antibiotics include penicillin, aminoglycosides, quinolones, and cephalosporins.

- Select antibiotics on the basis of surrounding and sputum smear Gram stain to do the empiric treatment.
- Gram stain positive pathogens are predominant in community-acquired infection mostly, while Gram stain negative pathogens are predominant in hospital-acquired infection.

Treatment

Compensation:

- Breath training
- Elevate the power of resistance
- Improve nutritional status
- Home oxygen therapy

Long term oxygen therapy is indicated for patients with persistent arterial hypoxemia at rest or after exercise (arterial oxygen tension consistently below 55mmHg while breathing room air.)

Preventive measure:

- Prevent respiratory infection
- Physical exercise
- Environmental health
- Stop smoking
- Lung function monitoring
**Acute cor pulmonale**

- Treatment of pulmonary embolism
- Cautious expansion of blood volume to maintain cardiac output
- Inhalation of 100% oxygen
- Primary therapy
  - Clot dissolution with thrombolysis or
  - Removal of pulmonary embolus by embolectomy
- Secondary prevention
  - Anticoagulation with heparin and warfarin and/or
  - Placement of an inferior vena cava filter

**Summary**

Cor Pulmonale

- Is an end stage manifestation of primary right sided heart failure.
- For the most part, treatment is supportive.
- In COPD, oxygen is a mainstay of therapy.
- Diuretics, ACEI, ARB, beta blockers may add efficacy.
- Better drug therapy, directed at pulmonary artery relaxation, may be on the horizon.
- Whatever the etiology the prognosis remains poor

**Thank you**