Supportive module 3 "Basics of diagnosis, treatment and prevention of major pulmonary diseases "

# Respiratory Failure & Cor Pulmonale

LECTURE IN INTERNAL MEDICINE FOR IV COURSE STUDENTS

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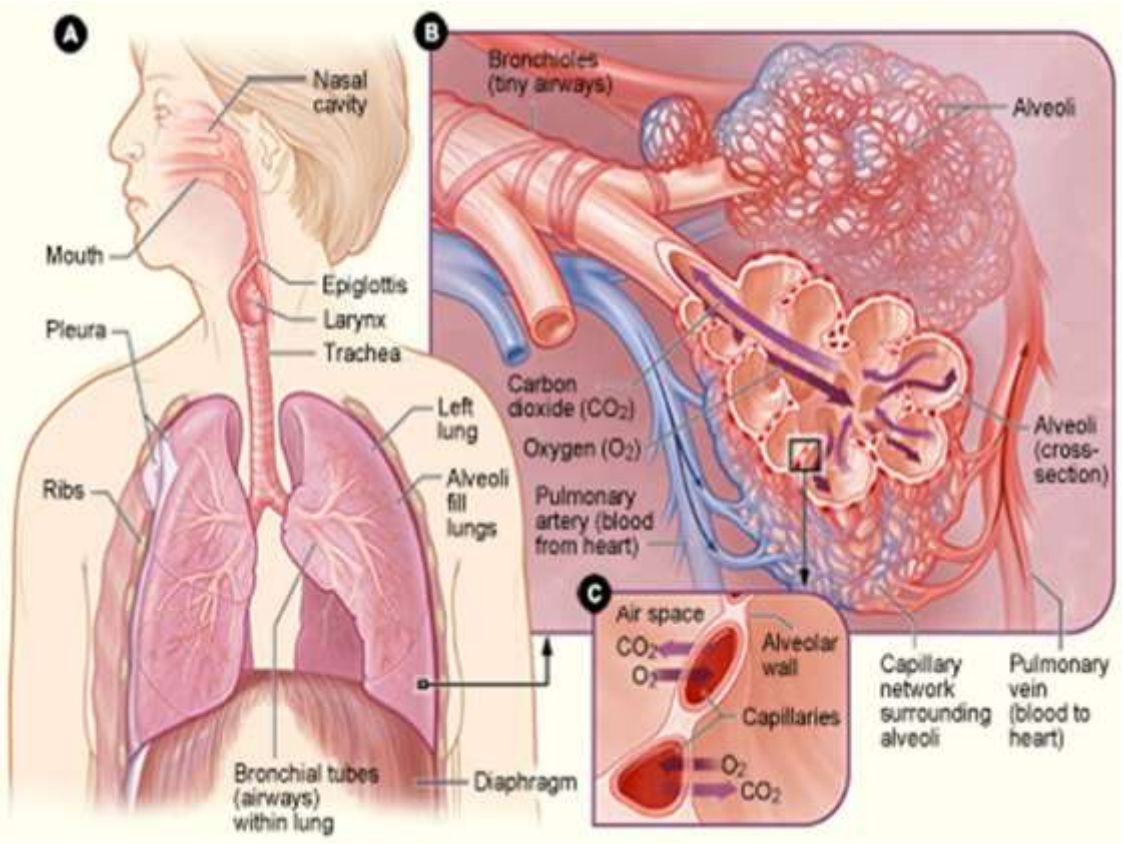
# Plan of the Lecture

## Respiratory Failure




- Definition
- Epidemiology
- Risk Factors and Etiology
- Mechanisms
- Classification
- Clinical presentation
- Diagnosis
- Treatment
- Prognosis
- Prophylaxis
- Abbreviations
- Diagnostic guidelines

http://images.slideplayer.com/26/8464013/slides/slide_5.jpg

# Respiratory Failure

# Definition

A 50-year-old male presents to the emergency department with sharp epigastric pain of 7 hours duration. The pain was sudden in onset and radiates to the back. The patient has a history of chronic alcoholism. The patient is admitted to the hospital and given 1 liter of normal saline. Several hours later, the patient appears markedly short of breath. T is 36.9 degrees Celsius, BP is 130/75 mmHg, pulse is 110/min, and RR is 33/min. Physical examination is notable for labored breathing and crackles at both lung bases. Which of the following would help confirm a diagnosis of Acute Respiratory Distress Syndrome (ARDS)?

1. White blood cell (WBC) count > 18,000 cells/mm^3
2. Blood glucose > 220 mg/dL, 3. Serum lactate dehydrogenase (LDH) > 400, 4. $PaO_2$ < 60 mmHg, 5. Pulmonary capillary wedge pressure (PCWP) < 18 mmHg.

http://emedicine.medscape.com/article/167981-overview en.wikipedia.org/wiki/Respiratory_failure

# USMLE TEST

The correct answer is 5: This patient has acute pancreatitis, and later experiences symptoms of respiratory failure consistent with ARDS. A PCWP < 18 mmHg would provide additional clinical evidence that the respiratory failure was secondary to ARDS and not hydrostatic edema.

Incorrect answers:
1-3: White blood cell count, blood glucose, and serum LDH are 3 of the 5 Ranson's criteria for pancreatitis upon hospital admission. The others are age > 55 years and serum AST > 250 IU/L. Ranson's criteria predict the severity of acute pancreatitis but do not contribute to diagnosis of ARDS.
4: See above.

https://wire.ama-assn.org/education/usmle-step-2-prep-next-best-step-hypertension-and-shortness-breath

# Epidemiology

- Incidence: about 360,000 cases per year in the United States
- 36% die during hospitalization
- Morbidity and mortality rates increase with age and presence of comorbidities and presence of comorbidities.

https://www.thoracic.org/professionals/clinical-resources/critical-care/clinical-education/mechanical-ventilation/respiratory-failure-mechanical-ventilation.pdf

# Risk Factors and Etiology

- A variety of pharmacologic, structural, and metabolic disorders of the CNS are characterized by depression of the neural drive to breathe
- Disorders of the peripheral nervous system, respiratory muscles, and chest wall lead to an inability to maintain a level of minute ventilation appropriate for the rate of carbon dioxide production
- Severe airway obstruction is a common cause of acute and chronic hypercapnia
- Diseases of the alveoli are characterized by diffuse alveolar filling, frequently resulting in hypoxemic respiratory failure, although hypercapnia may complicate the clinical picture.

http://emedicine.medscape.com/article/167981-overview#a5

# Risk Factors and Etiology

## Type I RF

- Chronic obstructive pulmonary disease (COPD)
- Pneumonia
- Pulmonary edema
- Pulmonary fibrosis
- Asthma
- Pneumothorax
- Pulmonary embolism
- Pulmonary arterial hypertension
- Pneumoconiosis
- Granulomatous lung diseases
- Cyanotic congenital heart disease
- Bronchiectasis
- Acute respiratory distress syndrome (ARDS)
- Fat embolism syndrome
- Kyphoscoliosis
- Obesity

http://emedicine.medscape.com/article/167981-overview#a5

# Risk Factors and Etiology

## Type II RF

- COPD
- Severe asthma
- Drug overdose
- Poisonings
- Myasthenia gravis
- Polyneuropathy
- Poliomyelitis
- Primary muscle disorders
- Porphyria
- Cervical cordotomy
- Head and cervical cord injury
- Primary alveolar hypoventilation
- Obesity-hypoventilation syndrome
- Pulmonary edema
- ARDS
- Myxedema
- Tetanus

http://emedicine.medscape.com/article/167981-overview#a5

# Mechanism

- RF may result from either a reduction in ventilatory capacity or an increase in ventilatory demand (or both)
- Ventilatory capacity can be decreased by a disease process involving any of the functional components of the respiratory system and its controller
- Ventilatory demand is augmented by an increase in minute ventilation and/or an increase in the work of breathing.

http://emedicine.medscape.com/article/167981-overview#a4

# Mechanism

## Hypoxemic (type I) RF

- The mechanisms are ventilation-to-perfusion ratio (V/Q) mismatch and shunt
- The low-V/Q units contribute to hypoxemia and hypercapnia and may occur either from a decrease in ventilation secondary to airway or interstitial lung disease or from overperfusion in the presence of normal ventilation
- Shunt is defined as the persistence of hypoxemia despite 100% oxygen inhalation; the deoxygenated blood (mixed venous blood) bypasses the ventilated alveoli and mixes with oxygenated blood that has flowed through the ventilated alveoli, consequently leading to a reduction in arterial blood content.

http://emedicine.medscape.com/article/167981-overview#a4

# Mechanism

## Hypercapnic (type II) respiratory failure

- At a constant rate of carbon dioxide production, $PaCO_2$ is determined by the level of alveolar ventilation and the relation between them is hyperbolic
- As ventilation decreases below 4-6 L/min, $PaCO_2$ rises precipitously
- A reduction in minute ventilation is observed primarily in the setting of neuromuscular disorders and CNS depression
- Hypoventilation occurs from depression of the CNS from drugs or neuromuscular diseases affecting respiratory muscles
- Hypoventilation is characterized by hypercapnia and hypoxemia.

http://emedicine.medscape.com/article/167981-overview#a4

# Mechanism

## Etiologic Categories

Type I: alveolar unit failure (collapse , flooding (edema, blood, pus, aspiration), fibrosis), pulmonary vasculature failure): pulmonary embolism, pulmonary hypertension

Type II: nervous system failure (central hypoventilation, neuropathies), muscle (pump) failure (muscular dystrophies, myopathies), neuromuscular transmission failure (myasthenia gravis), airway failure: obstruction, dysfunction), chest wall and pleural space failure (kyphoscoliosis, morbid obesity, pneumothorax , hydrothorax, hemothorax).

https://www.thoracic.org/professionals/clinical-resources/critical-care/clinical-education/mechanical-ventilation/respiratory-failure-mechanical-ventilation.pdf

# Classification

## International Classification of Diseases

- X Diseases of the respiratory system
- J95-J99 Other diseases of the respiratory system
- J96 Respiratory failure, not elsewhere classified
- J96.0 Acute respiratory failure
- J96.1 Chronic respiratory failure
- J96.9 Respiratory failure, unspecified.


http://apps.who.int/classifications/icd10/browse/2016/en#/J45

# Clinical classification

## Types

- Type I or hypoxemic
- Type II or hypercapnic
- Type III or perioperative
- Type IV in patients who are intubated and ventilated in the process of resuscitation for shock.

http://www.webmd.com/asthma/tc/classification-of-asthma-topic-overview

# Classification

## Terms

- Acute
- Chronic
- Acute on chronic (e.g.  acute exacerbation of advanced COPD).

http://www.webmd.com/asthma/tc/classification-of-asthma-topic-overview

# Symptoms and Signs

**Acute**

- An inability to breathe
- Bluish coloration in the skin, fingertips, or lips
- Restlessness
- Anxiety
- Confusion
- Altered consciousness
- Rapid, shallow breathing
- Racing heart
- Profuse sweating

http://www.healthline.com/health/chronic-respiratory-failure#Symptoms2healthline.com/health/acute-respiratory-failure#Symptoms5

# Symptoms and Signs

**Chronic**

- Difficulty breathing or shortness of breath, especially when active
- Coughing up mucous
- Wheezing
- Bluish tint to the skin, lips, or fingernails
- Rapid breathing
- Fatigue
- Anxiety
- Confusion.

http://www.healthline.com/health/chronic-respiratory-failure#Symptoms2healthline.com/health/acute-respiratory-failure#Symptoms5

# History

- The diagnosis RF begins with clinical suspicion of its presence
- Confirmation of the diagnosis is based on arterial blood gas analysis
- Evaluation of an underlying cause must be initiated early, in the presence of concurrent treatment for acute RF
- Cardiogenic pulmonary edema usually develops in the context of a history of left ventricular dysfunction or valvular heart disease
- Noncardiogenic edema occurs in typical clinical contexts, such as sepsis, trauma, aspiration, pneumonia, pancreatitis, drug toxicity, and multiple transfusions).

http://emedicine.medscape.com/article/167981-clinical

# Physical Examination 1

- Localized pulmonary findings reflecting the acute cause of hypoxemia, may be readily apparent
- In ARDS, the manifestations may be remote from the thorax, such as abdominal pain or long-bone fracture
- Neurologic manifestations include restlessness, anxiety, confusion, seizures, or coma
- Asterixis may be observed with severe hypercapnia
- Arrhythmias may result from hypoxemia and acidosis
- Cyanosis, a bluish color of skin and mucous membranes, indicates hypoxemia
- Dyspnea often accompanies respiratory failure

http://emedicine.medscape.com/article/167981-clinical#b2

# Physical Examination 2

- Excessive respiratory effort, vagal receptors, and chemical stimuli all may contribute to the sensation of dyspnea
- Confusion and somnolence may occur in respiratory failure
- Myoclonus and seizures may occur with severe hypoxemia
- Polycythemia is a complication of long-standing hypoxemia
- Pulmonary hypertension is present in chronic RF
- Alveolar hypoxemia potentiated by hypercapnia causes pulmonary arteriolar constriction
- The increased pulmonary vascular resistance increases afterload of the right ventricle, which may induce right ventricular failure.

http://emedicine.medscape.com/article/167981-clinical#b2

# Complications

- Pulmonary: embolism, nosocomial pneumonia, barotrauma, pulmonary fibrosis, complications secondary to the use of mechanical devices
- Cardiovascular: hypotension, reduced cardiac output, arrhythmia, endocarditis, myocardial infarction
- Gastrointestinal: hemorrhage, gastric distention, stress ulceration, ileus, diarrhea, and pneumoperitoneum
- Nosocomial infections: pneumonia, urinary tract infections, and catheter-related sepsis
- Renal failure
- Nutritional: result of enteral or parenteral nutrition.

http://emedicine.medscape.com/article/167981-clinical#b3

# Diagnosis

- Arterial blood gas analysis
- Chest X-ray
- Complete blood count (CBC)
- Renal function tests and liver function tests
- Serum creatine kinase and troponin I
- Thyroid function tests
- Spirometry
- Echocardiography
- Pulmonary function tests
- Electrocardiography (ECG)
- Right heart catheterization
- Pulmonary capillary wedge pressure.

http://patient.info/doctor/respiratory-failure

# Diagnosis


Acute Pulmonary Edema, Respiratory Failure, and LBBB: There is sinus tach with left bundle branch block (LBBB). There is excessively discordant ST elevation in leads V1 and V2. The highest ST/S ratio is in V1, with a ratio of 8/30 = 0.27, highly suggestive of LAD occlusion.

https://www.researchgate.net/publication/10770800_Respiratory_failure_in_elderly_patients

# Diagnosis

## Hypoxic Respiratory Failure Criteria

- Hypoxic RF is diagnosed in patients without chronic lung disease when there is falling pulse oximetry from 92% saturation or initial pulse oximetry <80% saturation on room air (severe respiratory failure is diagnosed when arterial blood gas shows PaO2 of <60 mmHg on room air)
- Patients with chronic lung disease may have low pulse oximetry readings and baseline PaO2 values of 50 mmHg, making worsening hypoxia difficult to recognize
- Decreases of 10% from baseline oxygenation can indicate impending respiratory failure in patients with chronic lung disease.

http://bestpractice.bmj.com/best-practice/monograph/853/diagnosis/criteria.html

# Diagnosis

## Hypercapnic Respiratory Failure Criteria

- Hypercapnic RF is diagnosed in patients without chronic lung disease when there is hypoxia and acute elevation of arterial $PaCO_2$ >45 to 50 mmHg and associated acidosis (pH <7.35)
- Patients with chronic lung disease can usually tolerate $PaCO_2$ levels of up to 80 mmHg with secondary renal compensation
- Increasing acidosis (decreasing pH) in these patients indicates respiratory failure.

http://bestpractice.bmj.com/best-practice/monograph/853/diagnosis/criteria.html

# Management

- The first objective in the management of RF is to reverse and/or prevent tissue hypoxia
- Hypercapnia unaccompanied by hypoxemia generally is well tolerated and probably is not a threat to organ function unless  the arterial blood pH falls below 7.2
- A patient with acute RF generally should be admitted to a respiratory care unit or intensive care unit (ICU)
- Most patients with chronic RF can be treated at home with oxygen supplementation and/or ventilatory assist devices along with therapy for their underlying disease

http://emedicine.medscape.com/article/167981-treatment#d9

# Management

Noninvasive ventilation in immunocompromised patients with acute respiratory failure.

https://www.hamilton-medical.com/de/Products/Mechanical-ventilators/HAMILTON-C3.html

# Management

- Extracorporeal membrane oxygenation (ECMO) may be more effective than conventional management for patients with severe but potentially reversible RF
- Assurance of an adequate airway with endotracheal intubation and ventilation are required in cases of severe RF (PaO2 less than 50 mmHg);  another indication is airway protection in patients with altered mental status
- Once the airway is secured, attention is turned toward correcting the underlying hypoxemia, the most life-threatening facet of acute RF.

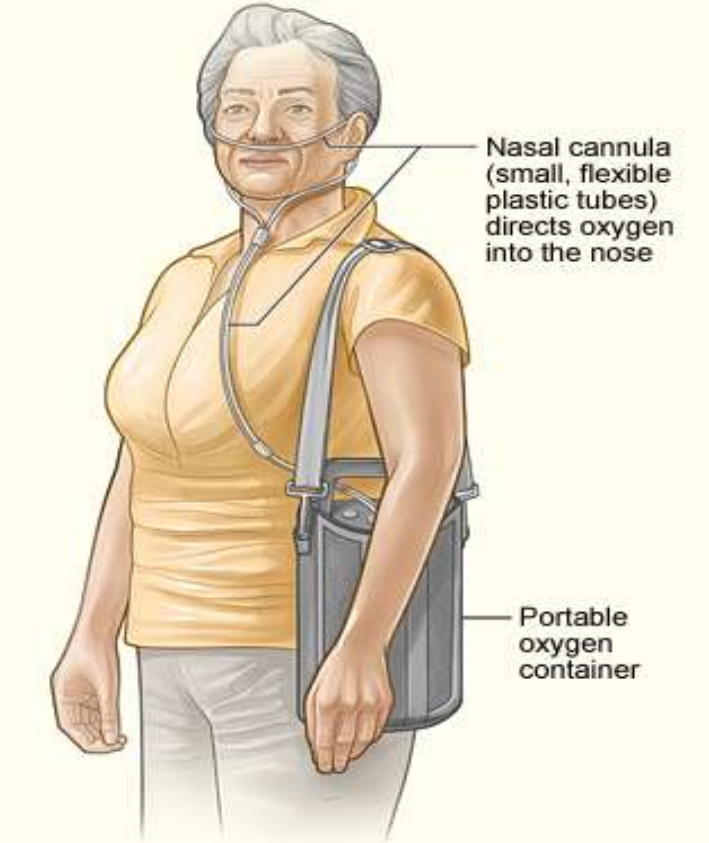
http://emedicine.medscape.com/article/167981-treatment#d9

# Management

- The goal is to assure adequate oxygen delivery to tissues, generally achieved with an arterial oxygen tension ($PaO_2$) of 60 mm Hg or an arterial oxygen saturation ($SaO_2$) greater than 90%
- Respiratory stimulants such as doxapram are rarely used, and if the RF resulted from an overdose of sedative drugs such as opioids or benzodiazepines, then the appropriate antidote (naloxone or flumazenil, respectively) will be given
- There is tentative evidence that in those with RF identified before arrival in hospital, continuous positive airway pressure can be useful when started before conveying to hospital.

https://en.wikipedia.org/wiki/Respiratory_failure

# Management




A nasal cannula and portable oxygen container are attached to a patient.

https://www.hamilton-medical.com/de/Products/Mechanical-ventilators/HAMILTON-C3.html

# Prognosis

- The mortality varies according to the etiology
- For ARDS, mortality is up to 45%
- Significant mortality occurs in patients admitted with hypercapnic RF
- For patients with COPD and acute RF, the overall mortality  is up to 26%.

http://emedicine.medscape.com/article/167981-overview#a7

# Prophylaxis

- Prevention is based on risk awareness of relevant medical conditions or trauma, and control or correction of underlying illnesses or injuries
- It is standard practice to administer influenza and pneumococcal vaccinations to patients at risk of RF with the thought that the vaccines control influenza and pneumococcal pneumonia and can help prevent RF in vulnerable groups
- Smoking cessation for all patients with lung disease limits the progression of pulmonary dysfunction
- Monitoring and continued medical management  helps limit acute exacerbations and reduces the risk of RF.

https://online.epocrates.com/diseases/85344/Acute-respiratory-failure/Prevention

# Abbreviations

ARDS - acute respiratory distress syndrome

CBC - complete blood count

CNS - central nervous system

COPD - chronic obstructive pulmonary disease

ECG - electrocardiography

ECMO - Extracorporeal membrane oxygenation

ICU - intensive care unit

RF - respiratory falure

TFTs - thyroid function tests

V/Q - ventilation-to-perfusion ratio.

# Diagnostic and treatment guidelines

Respiratory Failure

Treatment of respiratory failure in COPD

BTS/ICS guideline for the ventilatory management of acute hypercapnic respiratory failure in adults

Respiratory Failure Treatment & Management

Respiratory Failure

# Cor Pulmonale

# Plan of the Lecture

## Cor Pulmonale

- Definition
- Epidemiology
- Risk Factors  and Etiology
- Mechanisms
- Classification
- Clinical presentation
- Diagnosis
- Treatment
- Prognosis
- Prophylaxis
- Abbreviations
- Diagnostic guidelines

http://img.tfd.com/MosbyMD/thumb/cor-pulmonale.jpg

# Definition

Cor pulmonale (pulmonary heart disease, right hear failure) is the enlargement and failure of the right ventricle of the heart as a response to increased vascular resistance (such as from pulmonic stenosis) or high blood pressure in the lungs.

https://en.wikipedia.org/wiki/Pulmonary_heart_disease

# Definition

A 50-year-old male presents to the emergency department with sharp epigastric pain of 7 hours duration. The pain was sudden in onset and radiates to the back. The patient has a history of chronic alcoholism. The patient is admitted to the hospital and given 1 liter of normal saline. Several hours later, the patient appears markedly short of breath. T is 36.9 degrees Celsius, BP is 130/75 mmHg, pulse is 110/min, and RR is 33/min. Physical examination is notable for labored breathing and crackles at both lung bases. Which of the following would help confirm a diagnosis of Acute Respiratory Distress Syndrome (ARDS)?

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# USMLE TEST

The correct answer is 5: This patient has acute pancreatitis, and later experiences symptoms of respiratory failure consistent with ARDS. A PCWP < 18 mmHg would provide additional clinical evidence that the respiratory failure was secondary to ARDS and not hydrostatic edema.

Incorrect answers:
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4: See above.

https://wire.ama-assn.org/education/usmle-step-2-prep-next-best-step-hypertension-and-shortness-breath

# Epidemiology

- Cor pulmonale is estimated to account for 6-7% of all types of adult heart disease in the United States
- Cor pulmonale accounts for 10-30% of decompensated heart failure–related admissions in the United States
- Acute cor pulmonale is secondary to massive pulmonary embolism that is the most common cause of acute life-threatening cor pulmonale in adults (50,000 deaths in the United States)
- The incidence of cor pulmonale varies widely among countries, depending on the prevalence of cigarette smoking, air pollution, and other risk factors for various lung diseases.

http://emedicine.medscape.com/article/154062-overview?pa=yxfMHIhqouVY2QjSZ271hAHLllINrXw0FEuNAGQi9d45R6qiJxgtX19AtDT%2B5dxG8SIvl8zjYv73GUyW5rsbWA%3D%3D#a3

# Risk Factors and Etiology

- Acute respiratory distress syndrome (ARDS)
- Chronic obstructive pulmonary disease (COPD)
- Primary pulmonary hypertension
- Blood clots in lungs
- Kyphoscoliosis
- Interstitial lung disease
- Cystic fibrosis
- Sarcoidosis
- Obstructive sleep apnea (untreated)
- Sickle cell anemia
- Bronchopulmonary dysplasia (in infants).

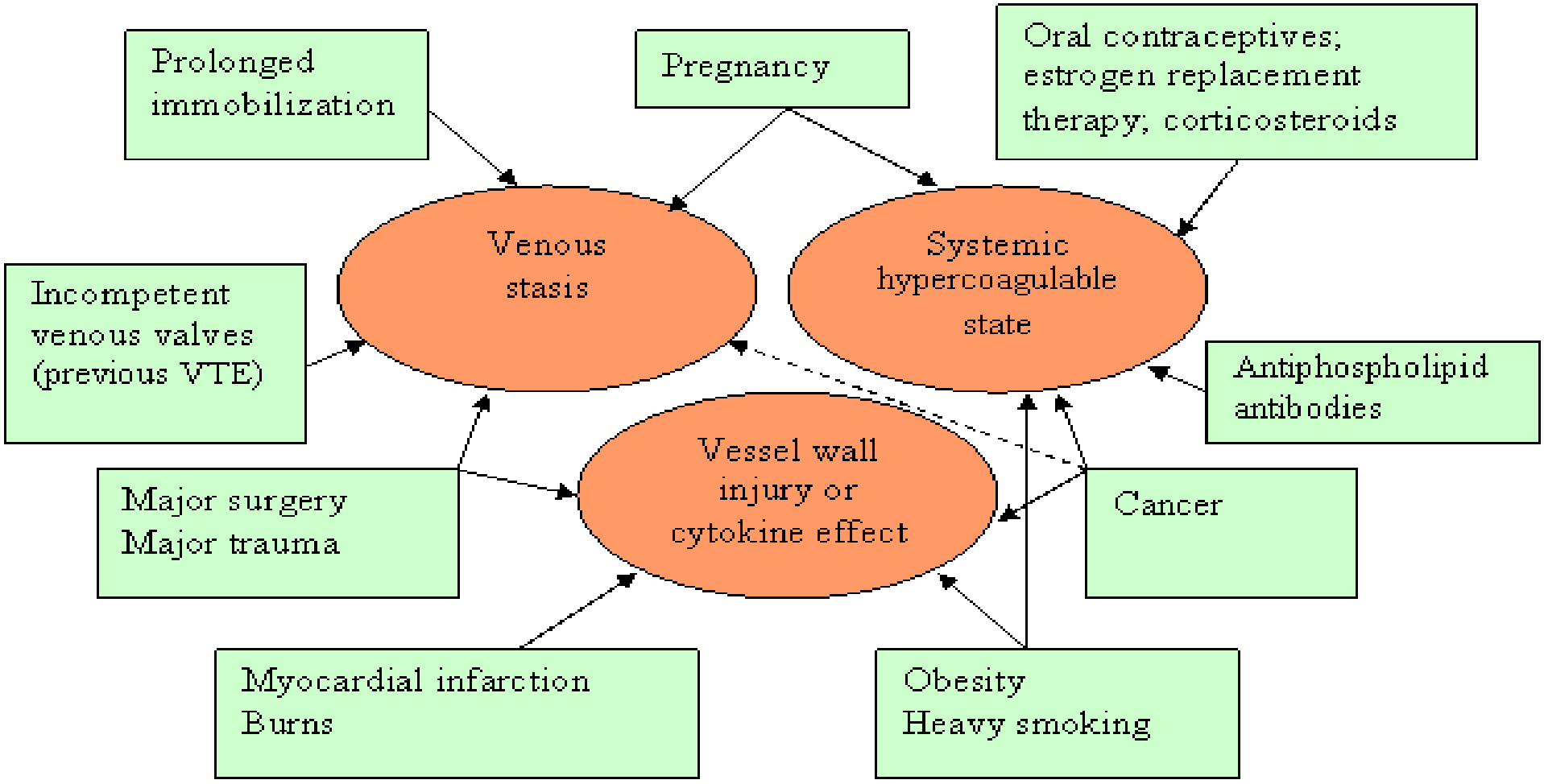
https://en.wikipedia.org/wiki/Pulmonary_heart_disease#Causes

# Mechanism

- Cor pulmonale characterised by the presence of peripheral edema, at least in some respiratory patients
- Acute cor pulmonale is result of pulmonary embolism (more common) and ARDS
- In ARDS, RV overload can occur due to mechanical ventilation and the pathologic features of the syndrome itself
- Mechanical ventilation, especially higher tidal volumes, requires a higher transpulmonary pressure
- In the case of ARDS, cor pulmonale is associated with an increased possibility of right-to-left shunting through a patent foramen ovale, which carries a poorer prognosis.

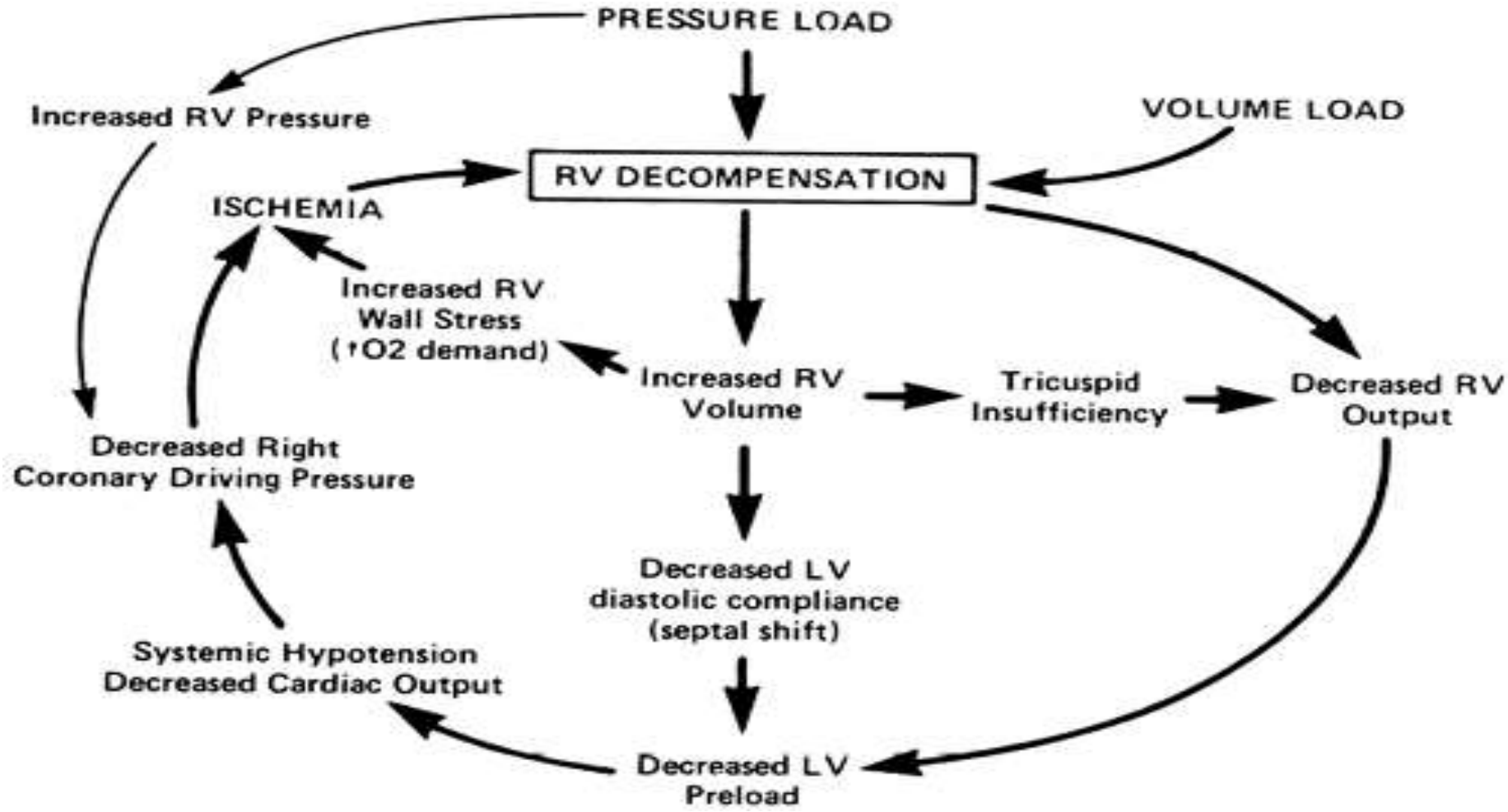
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1767533/

# Mechanism




Cor pulmonale and pulmonary embolism.

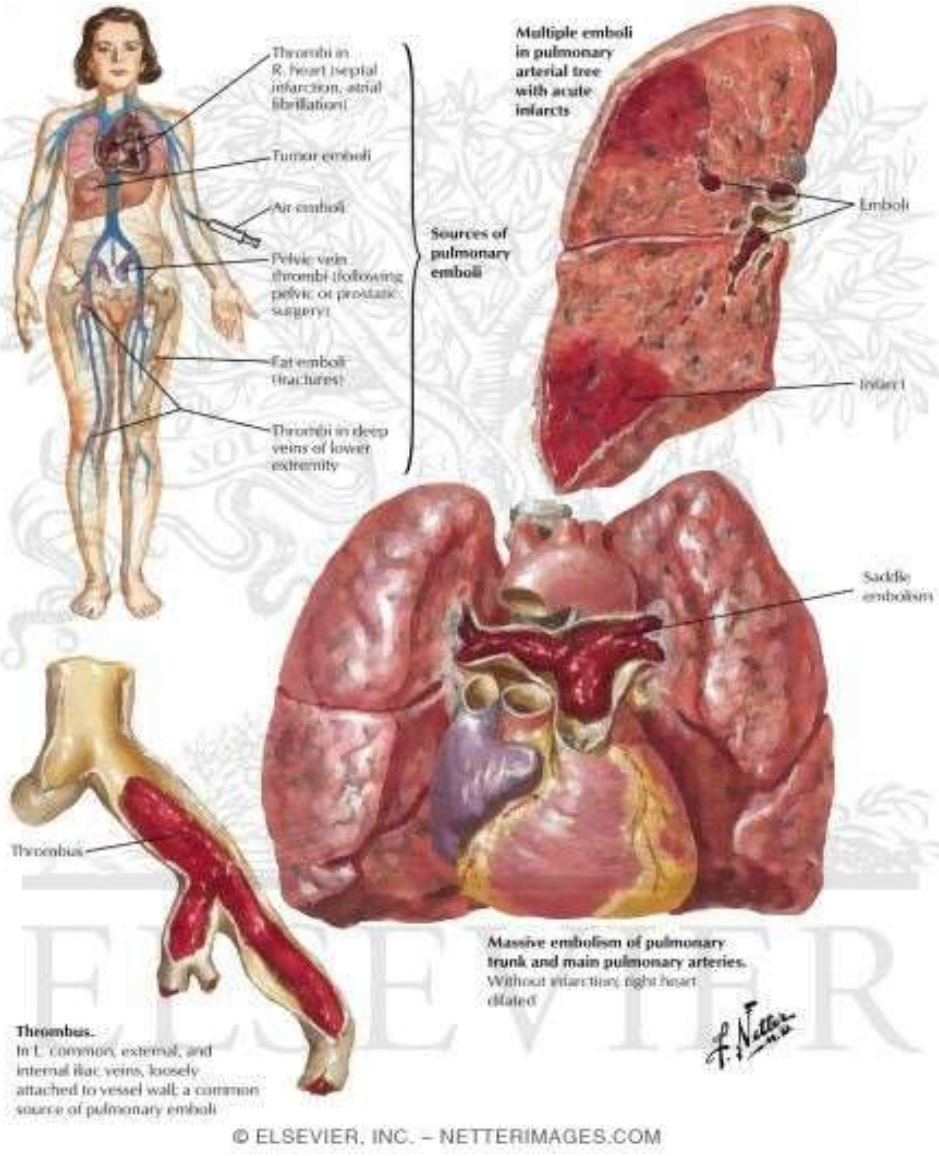
# Mechanism




Mechanism of acute right heart failure: the vicious cycle.

http://img.medscape.com/fullsize/migrated/458/659/src458659.fig4.gif

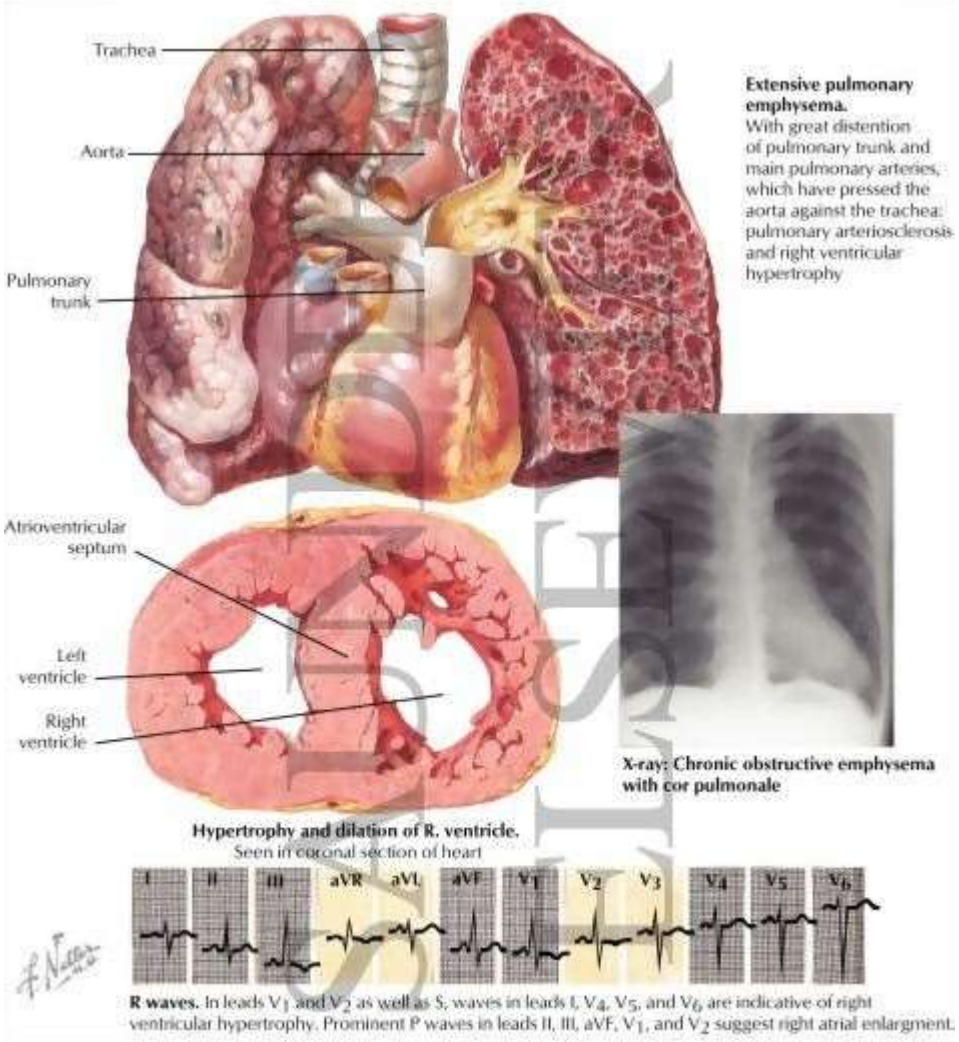
# Mechanism 7




Acute Cor Pulmonale.

https://www.netterimages.com/images/vpv/000/000/010/10281-0550x0475.jpg

# Mechanism 8





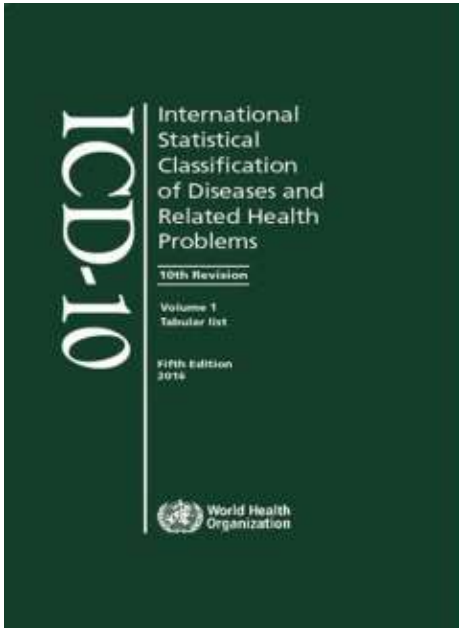

Chromic Cor Pulmonale.

https://www.netterimages.com/images/vpv/000/000/010/10281-0550x0475.jpg

# Classification

International Classification of Diseases .

- X Diseases of the respiratory system
- Pulmonary heart disease and diseases of pulmonary circulation (I26-I28)
- 126 Pulmonary embolism
- 127 Other pulmonary heart diseases
- 127.0 Primary pulmonary hypertension
- 127.1 Kyphoscoliotic heart disease
- 127.2 Other secondary pulmonary hypertension
- 127.8 Other specified pulmonary heart diseases
- 127.9 Pulmonary heart disease, unspecified.




http://apps.who.int/classifications/icd10/browse/2016/en#/J45

# Symptoms and Signs

- Shortness of breath
- Wheezing
- Cyanosis
- Ascites
- Jaundice
- Hepatomegaly
- Raised jugular venous pressure (JVP)
- Third heart sound
- Intercostal recession
- Presence of abnormal heart sounds
- Peripheral edema.

https://en.wikipedia.org/wiki/Pulmonary_heart_disease#Signs_and_symptoms

# Symptoms and Signs


https://s-media-cache-ak0.pinimg.com/originals/8c/b6/8b/8cb68b7c03b87a61906effb5eea9b4d8.jpg

# Clinical Manifestations

- The clinical manifestations of cor pulmonale may be nonspecific
- The symptoms may be subtle, especially in early stages of the disease, and they may be mistakenly attributed to the underlying pulmonary pathology
- Clinical signs are not sensitive indicators of pulmonary hypertension or right ventricular hypertrophy
- Peripheral edema is the best sign of right heart failure, but it is not specific and can arise from other causes.

# Complications

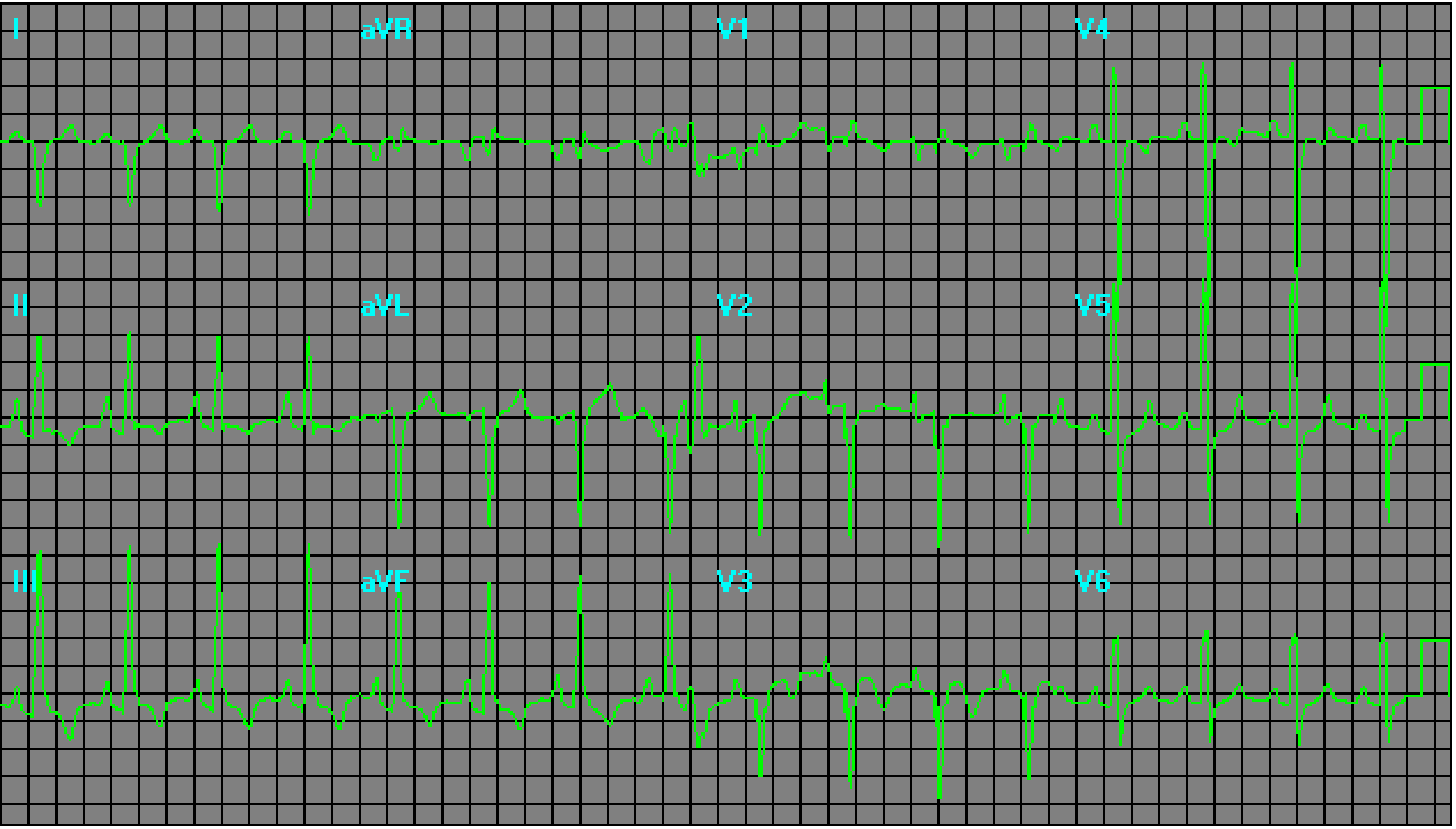
- Life-threatening shortness of breath
- Severe edema
- Shock
- Death.

https://medlineplus.gov/ency/article/000129.htm

# Diagnosis

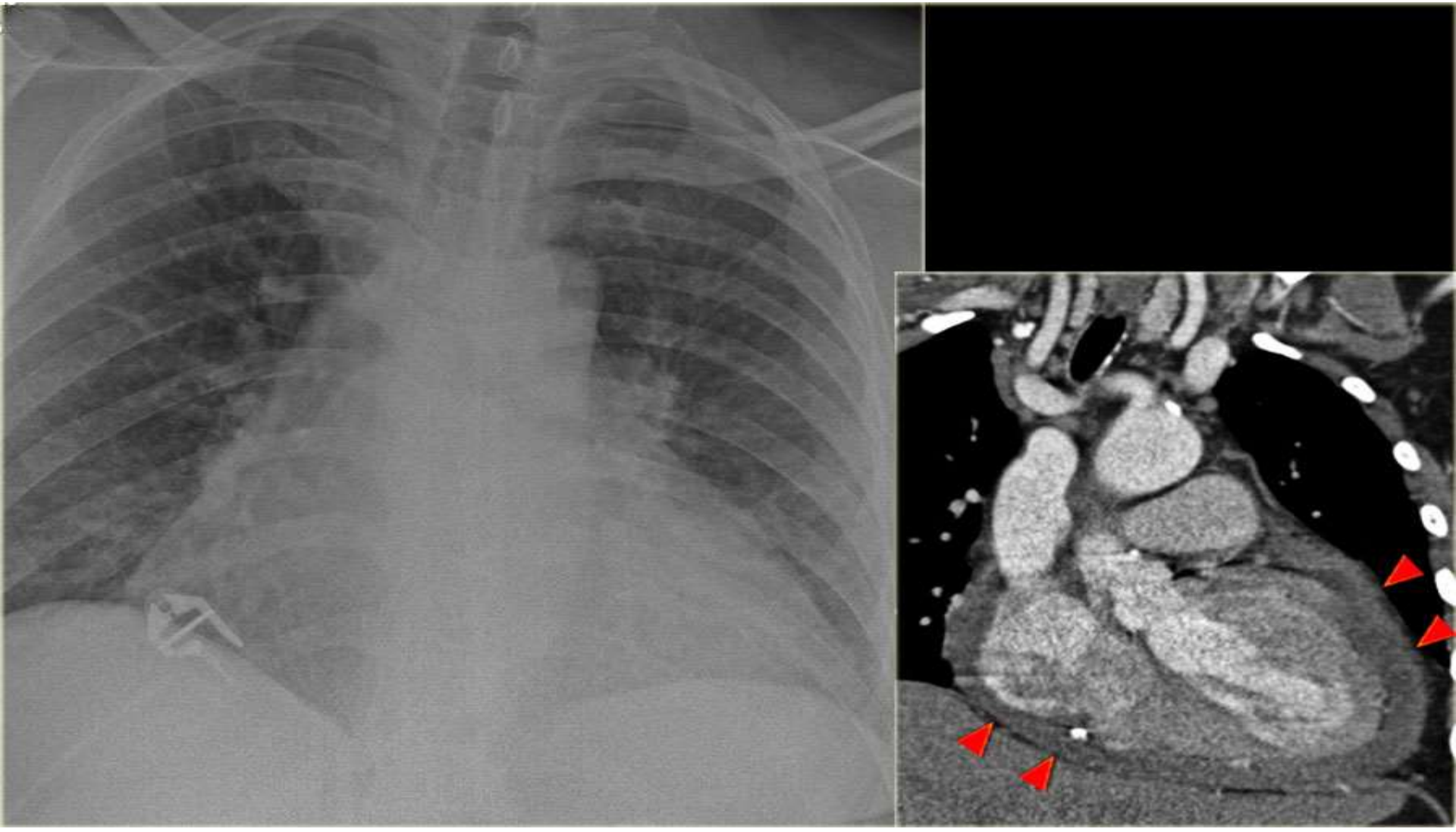
- Laboratory tests (hematocrit, serum alpha1-antitrypsin; antinuclear antibodies level for collagen vascular disease; and scleroderma; coagulations studies; plasma BNP, Arterial Blood Gas Analysis)
- Imaging studies (chest radiography, etc.), electrocardiography
- Right heart catheterization
- Pulmonary function tests
- Ventilation/perfusion  scanning

# Diagnosis



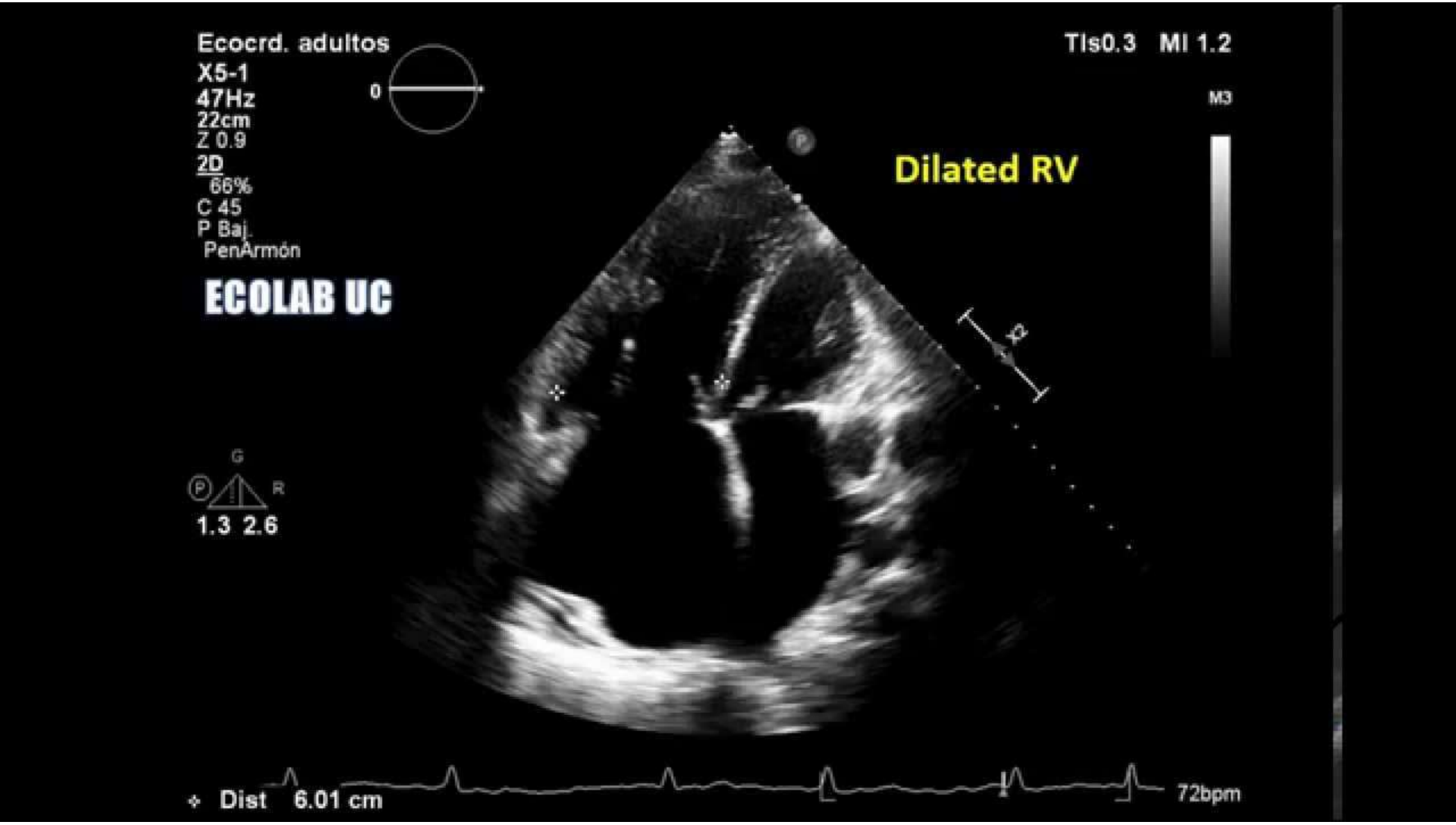

http://int-prop.lf2.cuni.cz/heart_sounds/ekg1/ekg-copd.gif

# Diagnosis



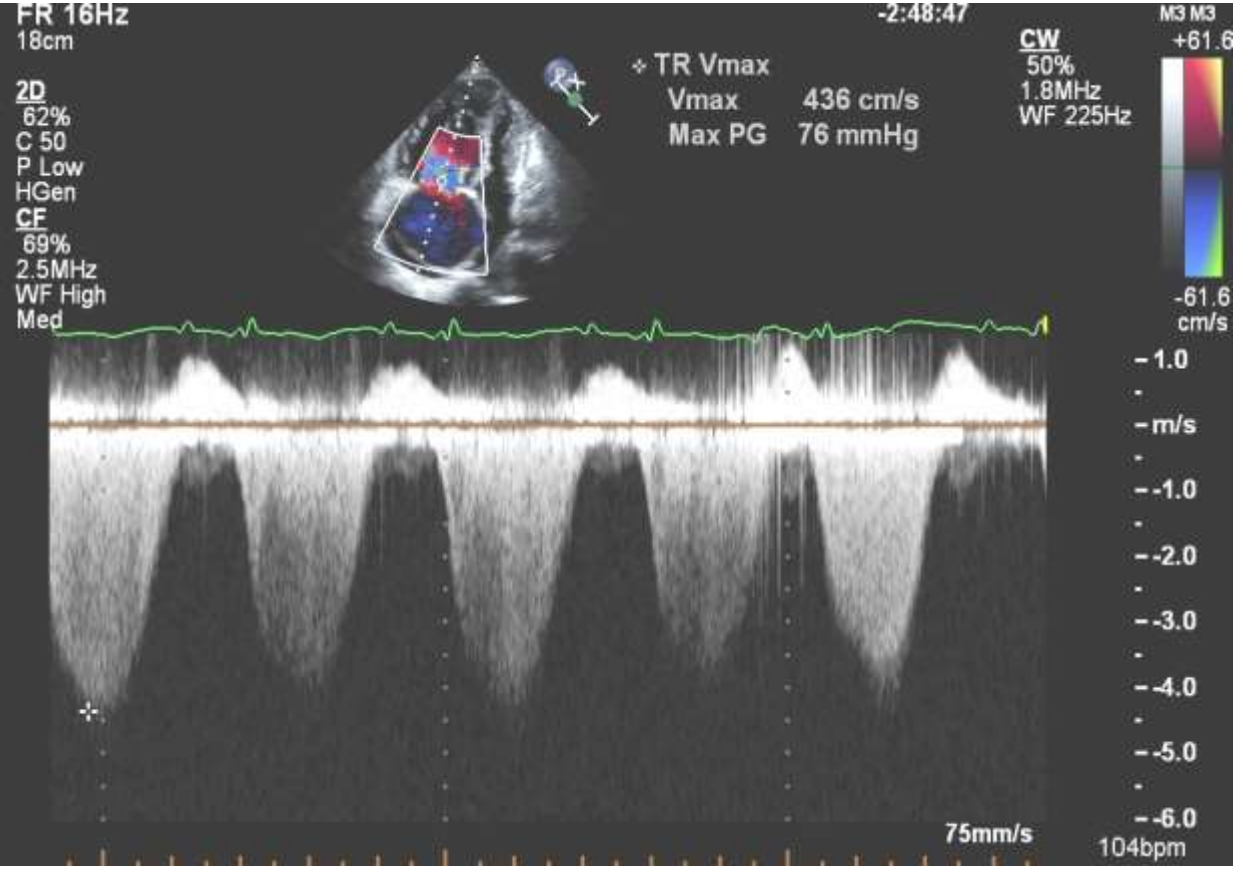
http://www.radiologyassistant.nl/data/bin/a509797a67fe0d_groot-cor-pericard.jpg

# Diagnosis




https://i.ytimg.com/vi/zaYxtKYthDk/maxresdefault.jpg

# Diagnosis




A thick dense regurgitated jet. The estimated pulmonary artery systolic pressure is markedly elevated.

https://i.ytimg.com/vi/zaYxtKYthDk/maxresdefault.jpg

# Diagnosis

Computerized axial tomography  demonstrating bilateral main pulmonary artery emboli.

https://i.ytimg.com/vi/zaYxtKYthDk/maxresdefault.jpg

# Diagnosis

## Differentiation

- Atrial myxoma
- Blood disorders associated with increased blood viscosity
- Congestive (biventricular) heart failure
- Constrictive pericarditis
- High-output heart failure
- Infiltrative cardiomyopathies
- Primary pulmonic stenosis
- Right heart failure due to right ventricular infarction and due to congenital heart diseases
- Ventricular septal defect.

# Management

## General

- Treatment of the underlying pulmonary disease and improving oxygenation and right ventricular function
- Oxygen therapy, diuretics, vasodilators, digitalis, theophylline, and anticoagulation therapy
- Cardiopulmonary support for acute cor pulmonale additionally includes fluid loading and vasoconstrictor (e.g., epinephrine) administration to maintain adequate blood pressure
- For massive pulmonary embolism in patients with acute cor pulmonale, consider administration of anticoagulation, thrombolytic agents or surgical embolectomy, especially if circulatory collapse is impending.

http://emedicine.medscape.com/article/154062-overview#a18

# Management

## Oxygen Therapy

- Oxygen therapy is of great importance, when administered on a continuous basis
- Although the impact of oxygen therapy on survival in patients with cor pulmonale is unclear, it may provide some degree of symptomatic relief and improvement in functional status
- Oxygen therapy plays an important role in the immediate setting and long-term management, especially in patients who are hypoxic and have COPD.

http://emedicine.medscape.com/article/154062-overview#a19

# Management

## Pharmacotherapy 1

- Diuretics are used when the right ventricle filling volume is markedly elevated and associated peripheral edema
- Vasodilators have been advocated in the long-term management of chronic cor pulmonale
- Calcium channel blockers (nifedipine, diltiazem), can lower pulmonary pressures
- Beta-selective agonists have an additional advantage of bronchodilator and mucociliary clearance effect
- Epoprostenol, treprostinil, and bosentan are prostacyclin analogues and have potent vasodilatory properties

http://emedicine.medscape.com/article/154062-overview#a20

# Management

## Pharmacotherapy 2

- The endothelin receptor antagonists are indicated in pulmonary artery hypertension
- The PDE5 inhibitors (sildenafil, tadalafil ) function by preventing the degradation of cyclic GMP and subsequently prolonging the vasodilatory effect of nitric oxide
- Riociguat (a soluble guanylate cyclase stimulant that mimics the function of nitric oxide as well as acts synergistically with it to promote vasodilation) has been FDA approved for the treatment of group I pulmonary hypertension as well as group 4 pulmonary hypertension (chronic thromboembolic pulmonary hypertension)

http://emedicine.medscape.com/article/154062-overview#a20

# Management

## Pharmacotherapy 3

- The use of cardiac glycosides, such as digitalis, must be used cautiously,  and should not be used during the acute phases of respiratory insufficiency
- Theophylline has been reported to reduce pulmonary vascular resistance and pulmonary arterial pressures acutely in patients with chronic cor pulmonale secondary to COPD
- Anticoagulation with warfarin is recommended in patients at high risk for thromboembolism
- Thrombolytic therapy is indicated in patients with acute cor pulmonale due to a pulmonary embolism resulting in hemodynamic instability.

http://emedicine.medscape.com/article/154062-overview#a20

# Management

## Surgical Approaches

- Phlebotomy is indicated in patients with severe polycythemia, defined as hematocrit of 65% or more
- Uvulopalatopharyngoplasty in selected patients with sleep apnea and hypoventilation
- Pulmonary embolectomy is indicated in patients with acute pulmonary embolism and hemodynamic instability when thrombolytic therapy is contraindicated, and in patients whose previous thrombolytic therapy failed
- Single-lung, double-lung, and heart-lung transplantation are all used to salvage the terminal phases of several diseases (e.g., emphysema, idiopathic pulmonary fibrosis, cystic fibrosis).

http://emedicine.medscape.com/article/154062-overview#a21

# Management

## Surgical Approaches 2

- Pulmonary embolectomy is indicated in patients with acute pulmonary embolism and hemodynamic instability when thrombolytic therapy is contraindicated, and in patients whose previous thrombolytic therapy failed, particularly if the location of the thrombus is in a more proximal location
- Single-lung, double-lung, and heart-lung transplantation are all used to salvage the terminal phases of several diseases (e.g., emphysema, idiopathic pulmonary fibrosis, cystic fibrosis).

http://emedicine.medscape.com/article/154062-overview#a21

# Management
## Outpatient Monitoring

- Patients with cor pulmonale generally require close attention in the outpatient setting
- It is appropriate to regularly assess the patient's oxygen needs and pulmonary function
- Consider a formal program of pulmonary rehabilitation, as many patients benefit from this therapy.

http://emedicine.medscape.com/article/154062-overview#a22

# Prognosis

- The occurrence of documented cor pulmonale is an indicator of poor prognosis in respiratory patients
- It is now accepted that a prolonged survival (≥ 10 years) can be observed after the first episode of peripheral edema
- The prevalence of clinical cor pulmonale has greatly decreased with the application of long term oxygen therapy, with a resulting improvement in prognosis.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1767533/

# Prophylaxis

Avoiding behaviors, such as cigarette smoking, which lead to chronic lung disease, may prevent the development of cor pulmonale.

http://www.nytimes.com/health/guides/disease/cor-pulmonale/overview.html

# Abbreviations

ARDS - acute respiratory distress syndrome

BNP - Brain natriuretic peptide

CBC - complete blood count

CNS - central nervous system

COPD - chronic obstructive pulmonary disease

HPV - hypoxic pulmonary vasoconstriction

ECMO - Extracorporeal membrane oxygenation

ICU - intensive care unit

JVP - jugular venous pressure

PAP - pulmonary arterial pressure

PVR - pulmonary vascular resistance

# Diagnostic and treatment guidelines

2015 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension

Cor pulmonale

Cor pulmonale

The treatment of chronic cor pulmonale

Pulmonary hypertension (guidelines on diagnosis and treatment of)

Diagnosis and management of stable chronic obstructive pulmonary disease