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# **PORTOPULMONARY HYPERTENSION AND HEPATOPULMONARY SYNDROME**

# PORTOPULMONARY HYPERTENSION

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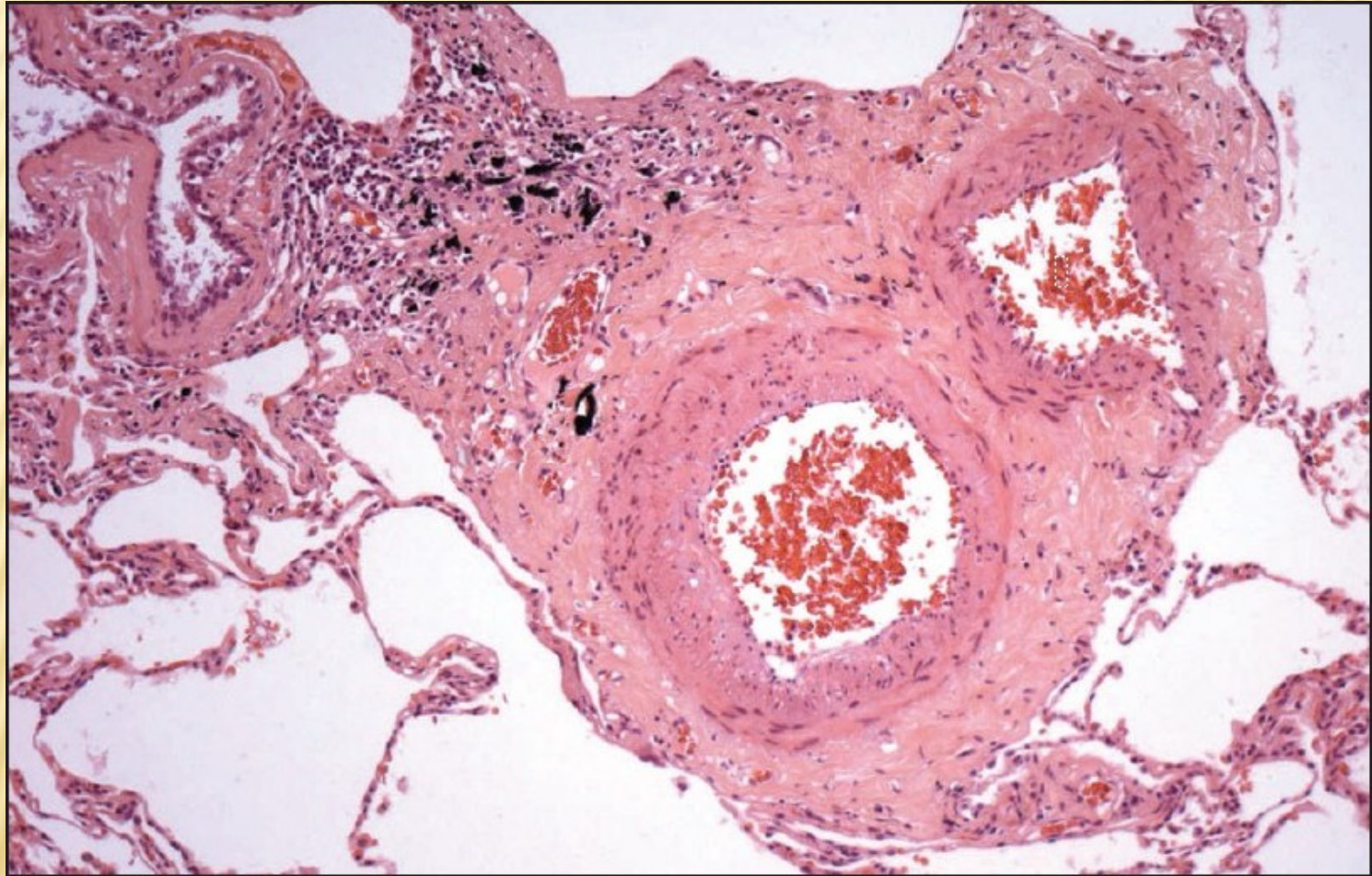
- ✘ Defined as the following:
  - + Raised Pulmonary Arterial Pressure
  - + Raised Pulmonary Vascular Resistance or a LVED Pressure of less than 15 mm Hg
  - + Occurring in the setting of Portal Hypertension
- ✘ Graded based on Pulmonary Arterial Pressure
  - + Mild 25-35 mm Hg
  - + Moderate 35-50 mm Hg
  - + Severe >50 mm Hg

# PORTOPULMONARY HYPERTENSION PATHOGENESIS

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- ✘ The development of portopulmonary hypertension seems to be independent of the cause of portal hypertension.
- ✘ The precise mechanism remains incompletely understood
  - ✘ Humoral Substance
  - ✘ Thromboembolism
  - ✘ Hyperdynamic Circulation

# PORTOPULMONARY HYPERTENSION HISTOLOGY



# PORTOPULMONARY HYPERTENSION

## CLINICAL FEATURES

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- ✗ Progressive Dyspnea on Exertion
- ✗ Fatigue
- ✗ Palpitations
- ✗ Syncope
- ✗ Chest Pain
- ✗ Jugular Venous Distention
- ✗ Loud Second Pulmonic Heart Sound of Tricuspid Regurgitation
- ✗ Lower Extremity Edema

# PORTOPULMONARY HYPERTENSION DIAGNOSIS

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- ✘ Clinical Suspicion
  - + Obtain a thorough History and Physical
- ✘ Transthoracic Echocardiogram
- ✘ V/Q Scan
- ✘ A Right Heart Catheterization is the Gold Standard

# PORTOPULMONARY HYPERTENSION MANAGEMENT

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- ✗ Diuretics for Volume Overload
- ✗ O<sub>2</sub> Supplementation for Hypoxemia
- ✗ Anticoagulation
- ✗ Prostacyclin Analogs
  - + Epoprostenol
- ✗ Oral Endothelin Receptor Antagonists
  - + Bosentan
- ✗ Phosphodiesterase-5 Inhibitors
  - + Sildenafil

# PORTOPULMONARY HYPERTENSION LIVER TRANSPLANTATION

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- ✘ Mild to moderate pulmonary hypertension is not associated with increased mortality post liver transplant
- ✘ Severe portopulmonary hypertension may be a contraindication to liver transplant given an associated high rate of mortality



# **THE HEPATOPULMONARY SYNDROME**

# HEPATOPULMONARY SYNDROME

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- ✘ Clinical syndrome with three components:
  - + Liver Disease
  - + Pulmonary Vascular Dilation
  - + Defect in Oxygenation
- ✘ Severity is measured based on the A-a Gradient

# DIAGNOSTIC CRITERIA FOR THE HEPATOPULMONARY SYNDROME

**Table 1. Diagnostic Criteria for the Hepatopulmonary Syndrome.\***

Variable	Criterion
Oxygenation defect	Partial pressure of oxygen <80 mm Hg or alveolar–arterial oxygen gradient ≥15 mm Hg while breathing ambient air
Pulmonary vascular dilatation	Positive findings on contrast-enhanced echocardiography or abnormal uptake in the brain (>6%) with radioactive lung-perfusion scanning
Liver disease	Portal hypertension (most common) with or without cirrhosis
Degree of severity†	
Mild	Alveolar–arterial oxygen gradient ≥15 mm Hg, partial pressure of oxygen ≥80 mm Hg
Moderate	Alveolar–arterial oxygen gradient ≥15 mm Hg, partial pressure of oxygen ≥60 to <80 mm Hg
Severe	Alveolar–arterial oxygen gradient ≥15 mm Hg, partial pressure of oxygen ≥50 to <60 mm Hg
Very severe	Alveolar–arterial oxygen gradient ≥15 mm Hg, partial pressure of oxygen <50 mm Hg (<300 mm Hg while the patient is breathing 100% oxygen)

\* All criteria were determined by means of positive contrast-enhanced echocardiography (i.e., microbubble opacification of the left heart chambers three to six cycles after right atrial passage). The abbreviated formula for the alveolar–arterial gradient is as follows:

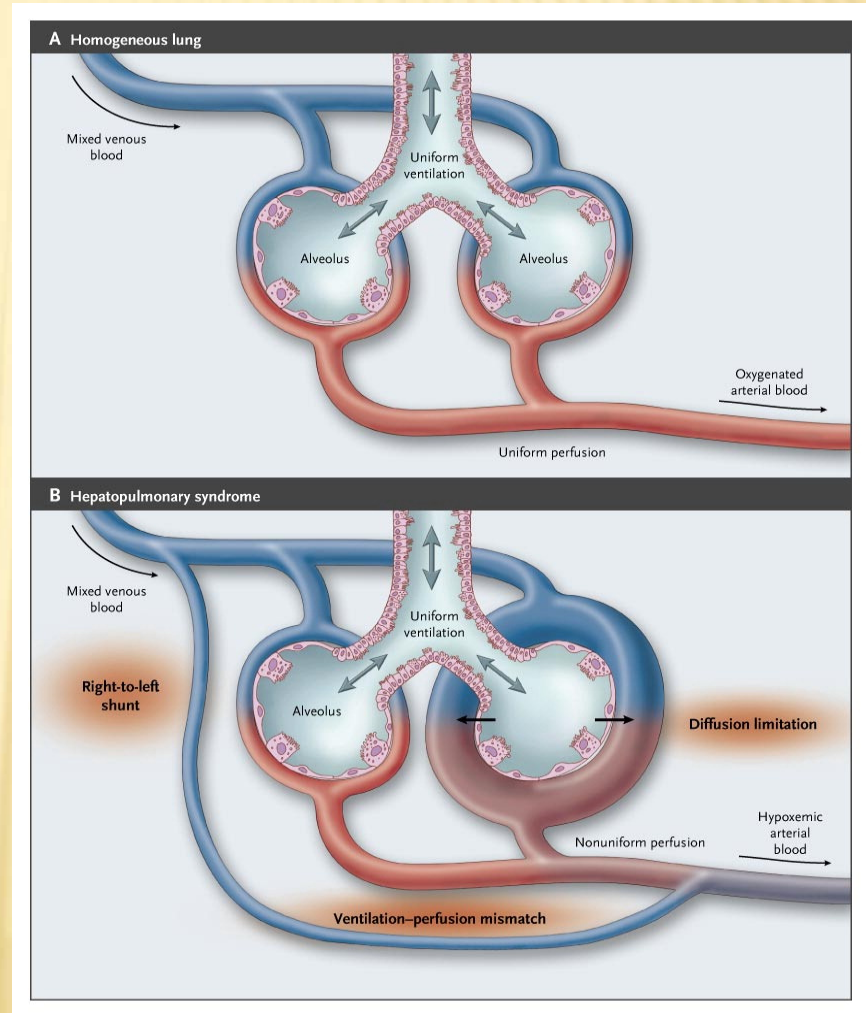
$$PAO_2 - PaO_2 = (F_I O_2 [P_{atm} - PH_2O] - [PaCO_2 / 0.8]) - PaO_2,$$

where  $PAO_2$  denotes partial pressure of alveolar oxygen,  $PaO_2$  partial pressure of arterial oxygen,  $F_I O_2$  fraction of inspired oxygen,  $P_{atm}$  atmospheric pressure,  $PH_2O$  partial pressure of water vapor at body temperature, and  $PaCO_2$  partial pressure of arterial carbon dioxide (0.8 corresponds to the standard gas-exchange respiratory ratio at rest); the normal range is 4 to 8 mm Hg (0.5 to 1.1 kPa). The normal range for the partial pressure of oxygen is 80 to 100 mm Hg (10.7 to 13.3 kPa) at sea level, while the patient is at rest and breathing ambient air. For patients older than 64 years of age, a value of ≤70 mm Hg (9.3 kPa) for  $PaO_2$  or ≥20 mm Hg for the alveolar-arterial gradient is often used. Ambient air is the respired gas unless otherwise indicated. To convert millimeters of mercury to kilopascals, multiply by 0.133.

† Data are from Rodríguez-Roisin et al.<sup>1</sup>

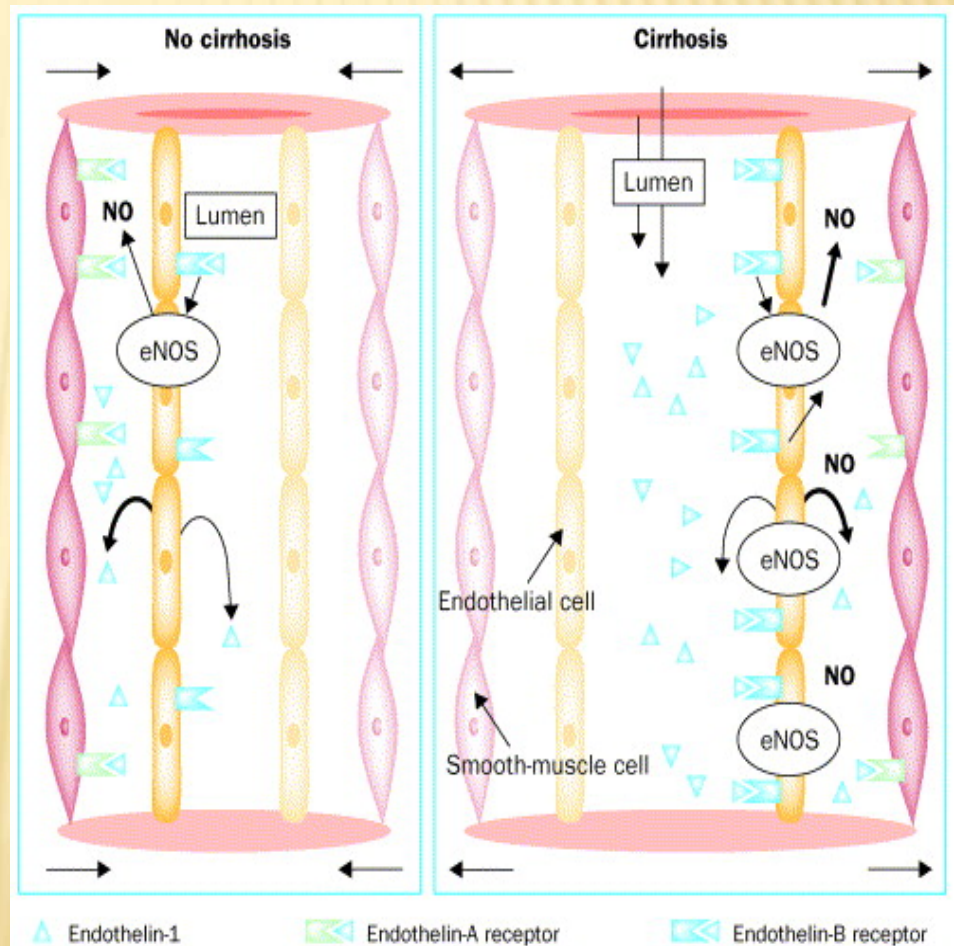
# HEPATOPULMONARY SYNDROME PATHOGENESIS

- ✘ Gross dilation of the pulmonary precapillary and capillary vessels
- ✘ Pleural and pulmonary arteriovenous communications
- ✘ Ventilation-perfusion mismatch



# HEPATOPULMONARY SYNDROME PATHOGENESIS

- ✘ Enhanced pulmonary production of NO has been implicated as a key priming factor for the development of pulmonary vascular dilation.
- ✘ Increased Endothelial Type B Endothelin Receptors in HPS lead to increased production of NO.



# HEPATOPULMONARY SYNDROME

## CLINICAL MANIFESTATIONS

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- ✘ Platypnea (dyspnea worsened by an erect position and improved by a supine position).
- ✘ Orthodeoxia (exacerbation of hypoxia and hypoxemia in an upright position)
- ✘ Progressive dyspnea
- ✘ Clubbing
- ✘ Distal cyanosis
- ✘ Spider Nevi

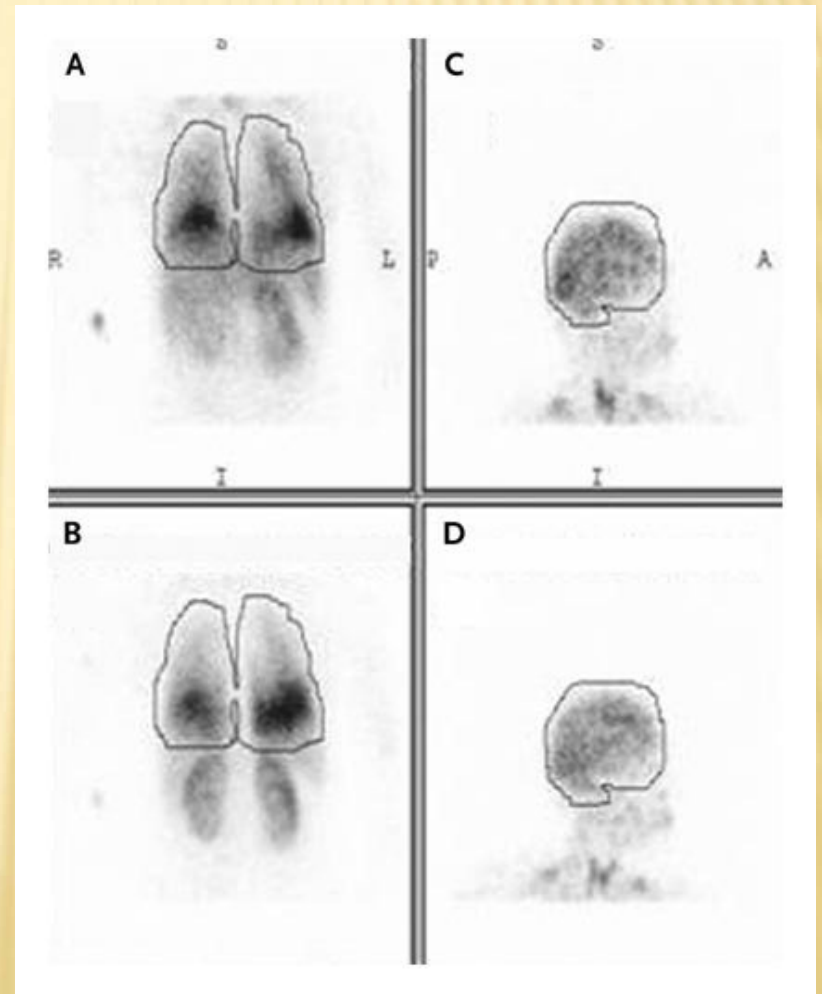
# HEPATOPULMONARY SYNDROME

## DIAGNOSIS

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- ✘ High degree of suspicion
- ✘ Measurement of ABG
- ✘ Detection of intrapulmonary shunting
  - + Contrast Echocardiography
  - +  $^{99m}\text{Tc}$  - labeled macroaggregated albumin scan
- ✘ Exclusion of intrinsic cardiopulmonary disease
  - + Chest CT
  - + PFT

# HEPATOPULMONARY SYNDROME DIAGNOSIS





# HEPATOPULMONARY SYNDROME

## TREATMENT

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- ✘ No effective medical therapies exist, and liver transplantation is the only successful treatment.
- ✘ Medical Therapy
  - + Garlic
  - + Trental
- ✘ TIPS

# HEPATOADRENAL SYNDROME

# ADRENAL INSUFFICIENCY IN LIVER DISEASE

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- ✘ Prevalence of RAI may range from 33% in severe liver failure to 69% in acute on chronic liver failure
- ✘ The current recommendations for patients with liver disease are to use corticosteroids in the presence of sepsis requiring vasopressors