شبكة المعلومات الجامعية

التوثيق الإلكتروني والميكروفيلم

بسم الله الرحمن الرحيم

HANAA ALY

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جامعة عين شمس

التوثيق الإلكتروني والميكروفيلم

قسم

نقسم بالله العظيم أن المادة التي تم توثيقها وتسجيلها
علي هذه الأقراص المدمجة قد أعدت دون أية تغيرات

يجب أن

تحفظ هذه الأقراص المدمجة بعيدا عن الغبار

HANAA ALY

Registry of Cases of Pulmonary Hypertension in Army Hospitals

Thesis

Submitted for Partial Fulfillment of MD in Chest Diseases

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List of Abbreviations

Abb.	Full term
5-HTT	5hydroxytryptamine transporter
6MWD	6-minute walking distance
6MWT	6-minute walking test
ABGs	Arterial blood gases
ADMA	Asymmetric dimethylarginine
BMP	Bone morphogenetic protein
BMPR	Bone morphogenetic protein receptor
BNP	Brain natriuretic peptide
BSA	Body surface area
cAMP	Cyclic adenosine monophosphate
CCB	Calcium channel blocker
cGMP	Cyclic guanosine monophosphate
CHD	Congenital heart diseases
CI	Cardiac index
CMPDs	Chronic myeloproliferative diseases
CMR	Cardiac magnetic resonance
CO	Cardiac output
COPD	Chronic obstructive pulmonary disease
CPET	Cardiopulmonary exercise testing
CRF	Chronic renal failure
CT	Computed tomography
CTD	Connective tissue disease
CTD	Connective tissue disease
CTEPH	Chronic thromboembolic pulmonary hypertension
CTEPH	Chronic thromboembolic pulmonary hypertension
DPAH	Drug-induced PAH

List of Abbreviations Cont...

Abb.	Full term
DPG	Diastolic pressure gradient
ECG	Electrocardiogram
EGF	Endothelial growth factor
EIF2AK4	Eukaryotic Translation initiation factor 2 alpha kinase 4
ERN	European Reference Networks
ERS	European Respiratory Society
ESC	European Society of Cardiology
ESC/ERS	European Society of Cardiology and the European Respiratory Society
ET-1	Endothelin-1
ETA	Endothelin A
ETB	Endothelin B
GD	Gaucher disease
GRACE	Good Research for Comparative Effectiveness
GSDs	Glycogen storage diseases
HFpEF	Heart failure with preserved ejection fraction
HIV	Human immunodeficiency virus
HPAH	Heritable pulmonary arterial hypertension
i.v.	Intravenous
IPAH	Idiopathic pulmonary arterial hypertension
KORPAH	Korean Registry of Pulmonary Arterial Hypertension
LV	Left ventricular
LVEDP	Left ventricular end-diastolic pressure
MAP	Mitogen-activated kinase
mPAP	Mean pulmonary arterial pressure
mSAP	Mean systemic arterial pressure
NHLBI	National Heart, Lung, and Blood Institute
NO	Nitric oxidase

List of Abbreviations Cont...

Abb.	Full term
NT-proBNP	N-terminal pro-brain natriuretic peptide
PA	Pulmonary artery
PAH	Pulmonary arterial hypertension
PAP	Pulmonary arterial pressure
PAPm	Mean pulmonary arterial pressure
PAWP	Pulmonary artery wedge pressure
PCA	Prostacyclin analogues
pCO2	Pressure of carbon dioxide
PDGF	Platelet-derived growth factor
PFT	Pulmonary function tests
PH	Pulmonary hypertension
PLCH	Pulmonary Langerhans cell histiocytosis
PPH	Primary PH
pred.	Predicted
PVOD	Pulmonary veno-occlusive disease
PVOD/PCH	Pulmonary veno-occlusive disease or pulmonary capillary hemangiomatosis
PVR	Pulmonary vascular resistance
PVRI	Pulmonary vascular resistance index
RA	Right atrium
RAP	Right atrial pressure
REVEAL	Registry to Evaluate Early And Long-term PAH disease management
RHC	Right heart catheterization
RV	Right ventricular
RVP	Right ventricular pressure
SAPH	Sarcoid-associated pulmonary hypertension
SCD	Sickle cell disease
SNAP	Surveillance of North American Pulmonary Hypertension

List of Abbreviations Cont...

Abb.	Full term
SOPHIA	Surveillance of Pulmonary Hypertension in America
SSc	Systemic sclerosis
SvO2	Mixed venous oxygen saturation
SVR	Systemic vascular resistance
TGF-b	Transforming growth factor-b
TPG	Transpulmonary pressure gradient
V/Q	Ventilation/perfusion
VE/VCO2	Ventilator equivalents for carbon dioxide
VEGF	Vascular endothelial growth factor
VIP	Vasoactive intestinal peptide
VO2	Oxygen consumption
VPAC	VIP receptors
WHO	World Health Organization
WHO-FC	World Health Organization functional class
WSPH	World Symposium on PH
WU	Wood units

INTRODUCTION

Pulmonary hypertension (PH) is a progressive disorder characterized by abnormally elevated blood pressure of the pulmonary circulation which results, over time, from extensive vascular remodeling and increased pulmonary vascular resistance (PVR)[1].

Pulmonary arterial hypertension (PAH) is a subcategory of pulmonary hypertension (PH) that comprises a group of disorders with similar pulmonary vascular pathology. Though PH is common, the estimated incidence of IPAH is 1–3 cases per million, making it a rare disease [1].

The hemodynamic definition of PAH is a mean pulmonary artery pressure at rest $\geq$ 25 mm Hg in the presence of a pulmonary capillary wedge pressure 15 $\leq$ mmHg with pulmonary vascular resistance (PVR) greater than 3 WU. Specific maneuvers during right heart catheterization can be utilized to disclose vasoreactivity and heart failure with preserved ejection fraction, which have implications for management [2].

Right heart catheterization is the most powerful maneuver for diagnosis confirmation by measuring the mean pressure of the pulmonary artery. It can also give more information about cardiac output and estimation of the pressure of the left atrium in order to assess the pressures of the left side