Introduction

Pulmonary hypertension (PH) has been defined as an increase in mean pulmonary arterial Pressure (MPAP) ≥25 mmHg at rest as assessed by right heart catheterization (RHC) ***(Nazzareno et al., 2009).***

It can be idiopathic (previously known as primary pulmonary hypertension (PPH)) or associated with other conditions or exposures, including connective tissue diseases, HIV infection, portal hypertension, and anorexigenic drug ingestion which is known as secondary pulmonary hypertension ***(Rubin, 2006).***

Manifestations of PH may not be apparent until pulmonary vascular disease is advanced, with symptoms of PH attributable to impaired oxygen transport and reduced cardiac output. Exertional dyspnea is the most frequent presenting symptom, with other complaints including fatigue, weakness, angina, syncope, peripheral edema, and abdominal distension ***(McGoon et al., 2004).***

Right heart catheterization is currently the gold standard method used to measure PH and to grade its severity. Cardiac catheterization, like any invasive procedure, is associated with important risks and complications, such as bleeding, infection, arrhythmias, or cerebrovascular events ***(Grubstein et al., 2008).***

Echocardiography is a key screening method in the diagnostic algorithm because it has special advantages over invasive procedures. It is safe, portable and repeatable. Therefore, echocardiography is more practical for evaluation of pulmonary artery hypertension (PAH) at the beginning of the disease and during the follow up ***(Ginghina et al., 2009).***

Computed tomography (CT) is commonly performed in patients suspected of having PH or in patients with an underlying diffuse lung disease who may be at risk for PH. In addition, the structure of the pulmonary vasculature at CT has been extensively studied as marker of increased MPAP. In this regard, most investigators have concentrated on dilation of the main pulmonary artery (both in absolute terms and in relation to the size of the ascending aorta) at axial CT as a sign of PH ***(Devaraj et al., 2008).***

Multi-detector CT angiography is now the first line of investigation in the diagnosis of most of pulmonary vascular disorders, it is a relatively available minimally invasive investigation that can now be considered as the first line of investigation in diagnosis of pulmonary embolism with its ability to detect small emboli down to the segmental and subsegmental and it is of value in patients with PH as it show different signs of PH as well as the possible cause ***(Addis et al., 2001).***

Aim of the Work

To evaluate correlation between pulmonary artery pressures measured by Doppler echocardiography and pulmonary arteries` diameters measured by CT angiography in patients with pulmonary hypertension secondary to respiratory disorders. Also to evaluate CT pulmonary angiography criteria of pulmonary hypertension secondary to respiratory disorders.

Pulmonary Vascular Anatomy

Pulmonary trunk arises from the base of the right ventricle (RV) above and to the left of the supra ventricular crest, then it slops up and back, at first in front of the ascending aorta, then to its left. Below the aortic arch, the pulmonary trunk divides at the level of the upper border of the fifth thoracic vertebra and to the left of the midline, into the right and left pulmonary arteries of almost equal size **(Fig.1)**.

The pulmonary trunk bifurcation lies below, in front and to the left of the tracheal bifurcation**,** which is also associated with the inferior tracheobronchial lymph nodes and the deep cardiac nerve plexus ***(Williams, 1995).***

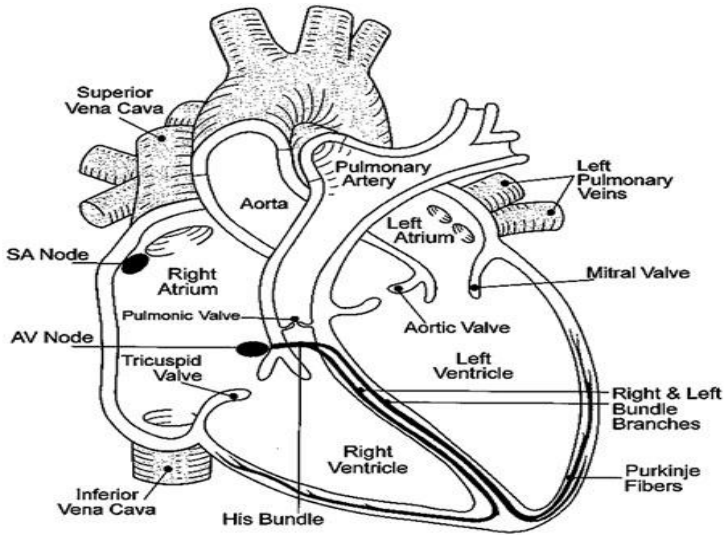

Figure (1): Diagram illustrate origin of pulmonary artery ***(Williams, 1995).***

The left pulmonary artery attached to the under surface of the aortic arch by the ligamentum arteriosum quickly spirals over the top of the left bronchus to reach the back of the bronchus and enter the hilum of the left lung. The right pulmonary artery, longer than the left passes below the carina anterior to the esophagus and at the lung root is held anterior to the right main bronchus by the upper lobe bronchus. It gives off its branch to the upper lobe and then enters the hilum ***(McMinn, 1994).***

Within the lung, each artery descends posterolateral to the main bronchus and divides into lobar and segmental arteries. Consequently, an arterial branch goes to each lobe and bronchopulmonary segment of the lung, usually on the anterior aspect of the corresponding bronchus. The arteries and bronchi are paired in the lung, branching simulatenously and running in parallel courses ***(Moore and Dalley, 1999).***

The pulmonary veins are short wide vessels passing to the left atrium (LA), usually two (upper and lower) from each lung. In the hilum of the lung, they lie below and in front of the artery. On the right, the upper passes behind the superior vena cava, the lower behind the right atrium (RA). On the left, both veins pass in front of the aorta ***(Lumley et al., 1995).***

The pulmonary veins carry well-oxygenated (arterial) blood from the lungs to the left atrium of the heart. Beginning in the pulmonary capillaries, the veins unite into larger and larger vessels. Inter segmental parts of pulmonary veins drain blood from adjacent bronchopulmonary segments into the inter-segmental parts of the pulmonary veins in the septa, which separate the segments. The pulmonary veins run independent courses from the arteries and bronchi as they run toward the hilum. The veins from the visceral pleura drain into the pulmonary veins, and the veins from the parietal pleura join the systemic veins in adjacent part of the thoracic wall ***(Moore and Dalley, 1999).***

The bronchial arteries have variable anatomy in terms of origin, branching pattern, and course ***(Yoon et al., 2002).***

They originate directly from the descending thoracic aorta, most commonly between the levels of the T5 and T6 vertebrae ***(Marshall and Jackson, 1997).***

It is reported that there is four classic bronchial artery branching patterns: two on the left and one on the right that presents as an intercostobronchial trunk (ICBT) (49.3% of cases); one on the left and one ICBT on the right (21%); two on the left and two on the right (one ICBT and one bronchial artery) (20%); one on the left and two on the right (one ICBT and one bronchial artery) (9.7%)

(Fig.2).The right ICBT is the most consistently seen vessel at angiography (80% of individuals). The right ICBT usually arises from the right posterolateral aspect of the thoracic aorta and the normal right and left bronchial arteries from the anterolateral aspect of the aorta. Right and left bronchial arteries that arise from the aorta as a common trunk are not uncommon at angiography. The true prevalence of a common bronchial artery trunk is unknown ***(Yoon et al., 2002).***

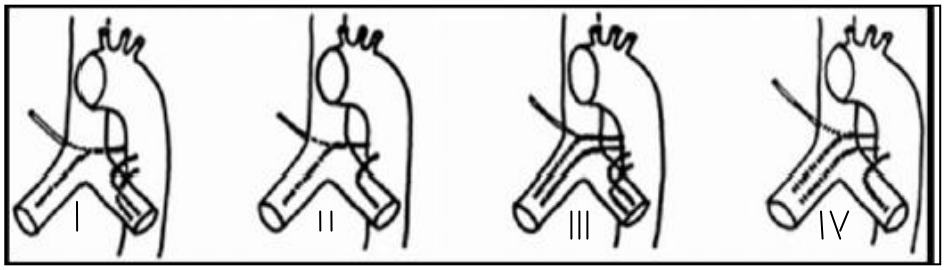

Figure (2): Diagrams illustrate the types of bronchial arterial supply: Type I, two bronchial arteries on the left and one on the right that manifests as an ICBT (49.3% of cases); Type II, one on the left and one ICBT on the right (21%); Type III, two on the left and two on the right (one ICBT and one bronchial artery) (20%); and Type IV, one on the left and two on the right (one ICBT and one bronchial artery) (9.7%) ***(Yoon et al., 2002).***

The bronchial arteries supply the trachea, extra- and intra- pulmonary airways, bronchovascular bundles, nerves, supporting structures, regional lymph nodes, visceral pleura and esophagus as well as the vasa vasorum of the aorta, pulmonary artery and pulmonary vein **(Fig.3)** ***(Deffenbach et al., 1987).***

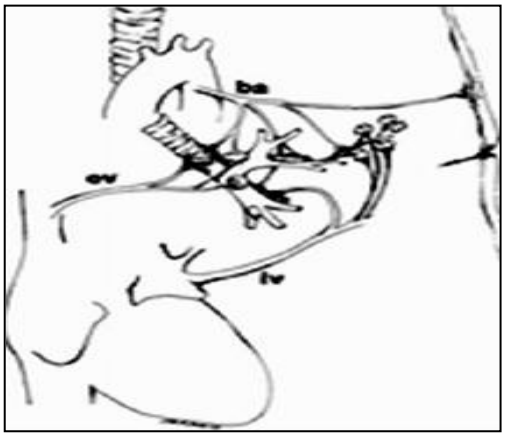

Figure (3): Schematic illustrates how the bronchial arteries **(ba)** supply the visceral pleura, airways, vasa vasora of pulmonary arteries, lymph nodes, and bronchovascular and neural bundles. Extra-pulmonary bronchial veins **(ev)** drain to the right side of the heart, and intrapulmonary veins **(iv)** anastomose with pulmonary arteries and return to the left side of the heart ***(Frazier et al., 2000).***

Each lung is typically supplied by two bronchial vessels that impart several branches to the extra-pulmonary mediastinal structures enroot to the pulmonary hila, where they enter the peribronchial sheath of each main stem airway. The bronchial arteries then form a dual-layered adventitial and sub-mucosal plexus along the airways that communicates freely across airway muscular walls and nourishes the bronchial tree down to the terminal bronchioles ***(Burke and Virmani, 1996).***

Normally, the bronchial circulation only supplies nutrients and is not involved in gas exchange. However, in certain pathologic conditions (e.g., occlusion of a main

pulmonary artery), the bronchial vessels do participate in blood oxygenation. The bronchial circulation responds with enlargement and hypertrophy to decreased pulmonary flow and ischemia **(Fig.4)**; trans-pleural systemic collateral vessels (eg, intercostals arteries, internal mammary arteries) also may develop ***(Yoon et al., 2002).***

The pulmonary and bronchial vascular networks communicate via several micro-vascular interconnections. There are also trans-pleural systemic–pulmonary artery anastomoses ***(Deffenbach et al., 1987).***

Unlike the pulmonary arteries, bronchial arteries are notably smaller than their adjacent airways and follow more tortuous paths ***(Burke and Virmani, 1996).***

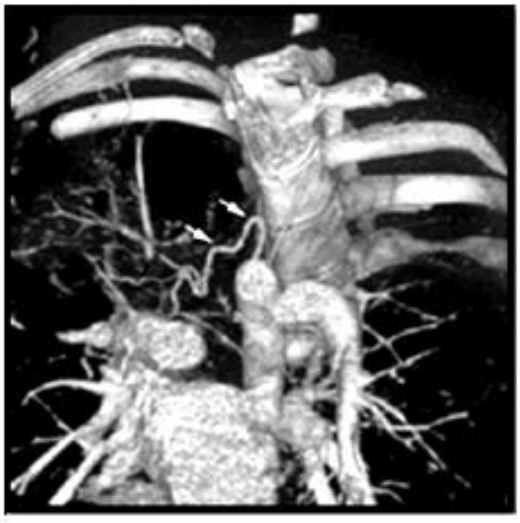

Figure (4): Volume-rendered CT image of thoracic vessels and posterior bone structures shows an enlarged right bronchial artery (arrows) originating from a right intercostobronchial trunk, with a tortuous mediastinal course **(yoon et al., 2002**)

Deep bronchial veins communicating freely with the pulmonary veins and eventually joining a single trunk which ends in a main pulmonary vein or in the left atrium. Superficial bronchial veins drain extra pulmonary bronchi, communicate with the pulmonary veins and end on the right in the azygous vein and on the left in the superior intercostals or the accessory hemi- azygous vein ***(Williams et al., 1995).***

PULMONARY VASCULAR HISTOLOGY

Pulmonary arteries main, lobar, segmental, and sub-segmental with a diameter greater than 0.5 mm are referred to as elastic pulmonary arteries ***(Castaner et al., 2006).***

The function of the elastic arteries is similar to that of the aorta, to provide a distensible reservoir for ventricular ejection. The normal pulmonary circulation is a low pressure system that has approximately one-tenth the flow resistance of the systemic circulation, as well as a high capacitance ***(Frazier et al., 2000).***

Beyond the sub-segmental bronchi, these vessels transition to muscular arteries which accompany the peripheral airways downward to the level of the terminal bronchioles. As the smooth-muscle layer progressively thins, these arteries become arterioles (0.15–0.015 mm in diameter), which proceed along the respiratory bronchioles and alveolar ducts to eventually form a capillary network in

the alveolar walls**.** Their walls comprise multiple parallel elastic lamellae, smooth muscle cells, and collagen fibrils. Medial smooth muscle fibers in the muscular arteries provide active vasodilatation and constriction ***(Frazier et al., 2000).***

The venules accept flow from these capillary beds and unite to form pulmonary veins, which course within interlobular fibrous septa, apart from the airways **(Fig.5)**. Two veins from each hilum drain into the left atrium ***(Frazier et al., 2000).***

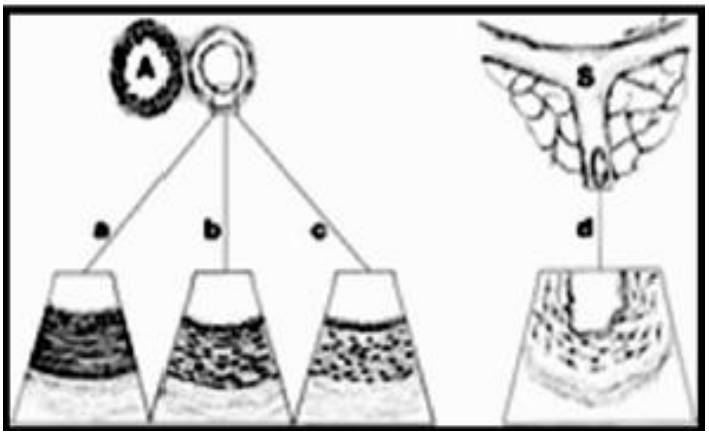

Figure (5): Cross-sectional drawings depict normal pulmonary vascular anatomy. Arterial vessels are located adjacent to a schematic airway **(A)**. The walls of elastic arteries **(a)** contain multiple parallel elastic lamellae, smooth muscle cells, and collagen fibrils. The muscular arteries **(b)** contain a media of smooth muscle fibers, bordered by distinct internal and external elastic laminae. Arterioles **(c)** are distinguished by the absence of a distinct external elastic lamina. Veins **(d)** are identified by their septal location **(S)** and a media of loosely organized smooth muscle fibers **(Frazier et al., 2000).**

Pulmonary Hypertension

Definition:

Pulmonary hypertension has been defined as an increase in mean pulmonary arterial Pressure (PAP) ≥ 25 mmHg at rest as assessed by right heart catheterization ***(Nazzareno et al., 2009).***

Recent re-evaluation of available data has shown that the normal mean PAP at rest is 14±3 mmHg, with an upper limit of normal of 20 mmHg, The significance of a mean PAP between 21 and 24 mmHg is unclear. Patients presenting with PAP in this range need further evaluation in epidemiological studies ***(Kovacs et al., 2009).***

The definition of PH on exercise as a mean PAP ≥30 mmHg as assessed by right heart catheterization is not supported by published data and healthy individuals can reach much higher values ***(kovacs et al., 2009).***

Classification of pulmonary hypertension:

It can be idiopathic pulmonary artery hypertension (IPAH) previously known as primary pulmonary hypertension or associated with other conditions or exposures, including connective tissue diseases, HIV infection, portal hypertension, and anorexigenic drug

ingestion which is known as secondary pulmonary artery hypertension (SPAH) ***(Rubin, 2006).***

During the fourth World Symposium on PH held in 2008 in Dana Point, California, the consensus agreement of experts worldwide was to maintain the general philosophy and organization of the Evian-Venice classifications while amending some specific points to improve clarity and to take into account new information. The new clinical classification (derived from the Dana Point meeting) is shown in the Table 1 ***(Simonneau et al., 2009).***

1 Pulmonary arterial hypertension

1.1 Idiopathic

1.2 Heritable

1.2.1 BMPR II (bone morphogenetic protein receptor, type II)

1.2.2 ALK1(activin receptor-like kinase 1 gene, endoglin (with or without hereditary hemorrhagic telangiectasia)

1.2.3 Unknown

1.3 Drugs and toxins induced

1.4 Associated with (APAH)

1.4.1 Connective tissue diseases

1.4.2 HIV infection

1.4.3 Portal hypertension

1.4.4 Congenital heart disease

1.4.5 Schistosomiasis

1.4.6 Chronic hemolytic anemia

1.5 Persistent pulmonary hypertension of the newborn

1.6 Pulmonary veno-occlusive disease(PVOD) and/or pulmonary capillary haemangiomatosis

2 Pulmonary hypertension due to left heart disease

2.1 Systolic dysfunction

2.2 Diastolic dysfunction

2.3 Valvular disease

3 Pulmonary hypertension due to lung diseases and/or Hypoxia.

3.1 Chronic obstructive pulmonary disease

3.2 Interstitial lung disease

3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern

3.4 Sleep-disordered breathing

3.5 Alveolar hypoventilation disorders

3.6 Chronic exposure to high altitude

3.7 Developmental abnormalities

4 Chronic thromboembolic pulmonary hypertension (CTEPH)
5 PH with unclear and/or multifactorial mechanisms. **5.1 Hematological disorders: myeloproliferative disorders, splenectomy.** **5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleio-myomatosis, neurofibromatosis, vasculitis** **5.3 Metabolic disorders: glycogen storage disease, Gaucher´s disease, thyroid disorders** **5.4 Others: tumoural obstruction, fibrosing mediastinitis, chronic Renal failure on dialysis**

(Simonneau et al., 2009)

Pathology of pulmonary hypertension:

Different pathological features characterize the diverse clinical PH groups.

Group 1, PAH: pathological lesions affect the distal pulmonary arteries (0.5 mm of diameter) in particular. They are characterized by medial hypertrophy, intimal proliferative and fibrotic changes (concentric, eccentric), adventitial thickening with moderate perivascular inflammatory infiltrates, complex lesions (plexiform,