INTERSTITIAL (DIFFUSE PARENCHYMAL) LUNG DISEASES IN PEDIATRICS

ESSAY
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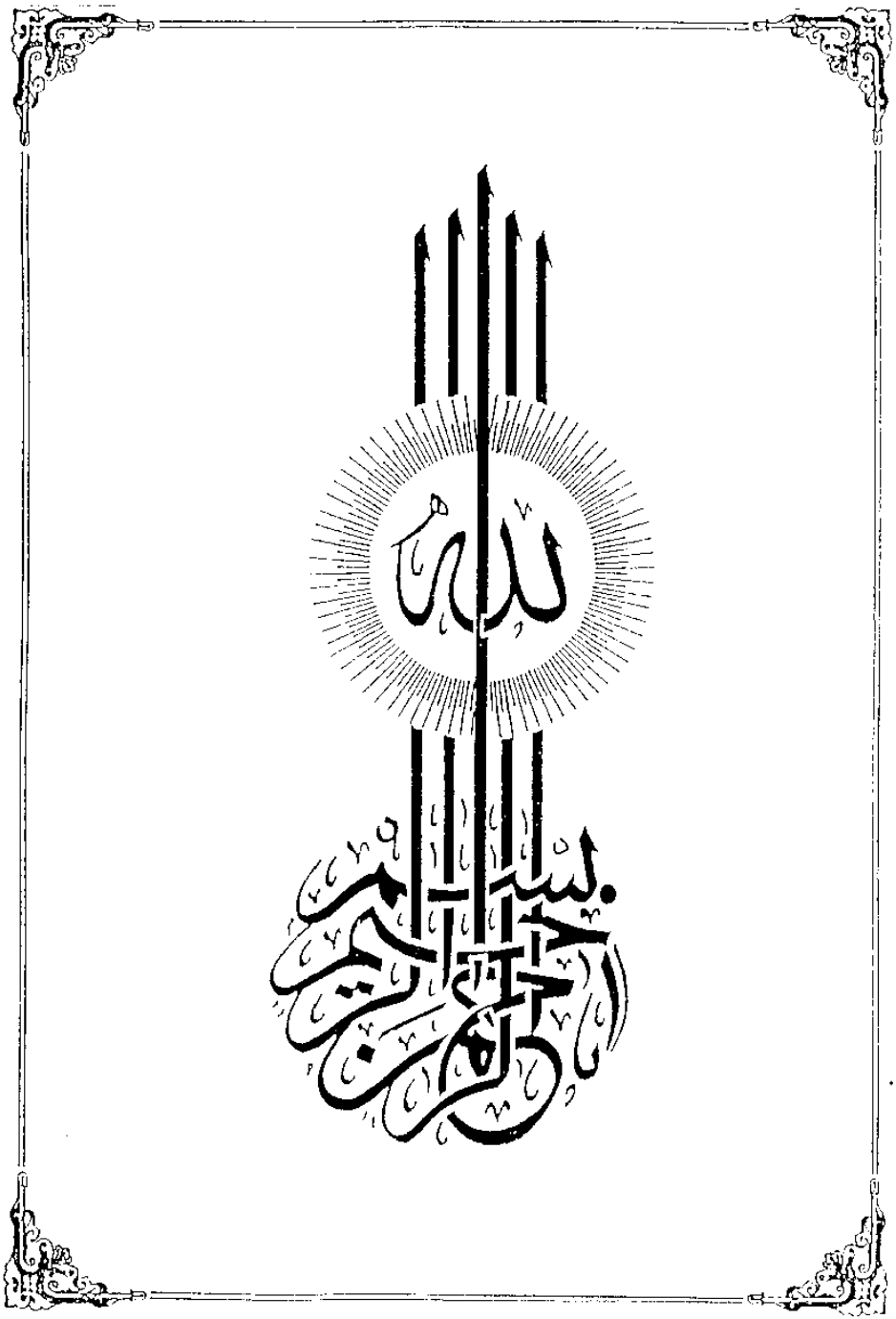

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ABBRIVIATION

Words	Abbriviate liters
Interstitial lung diseases	ILDs
Interstitial lung disease	ILD
Interstitial lung disease in children	ILD-C
Tidal volume	TV
Fraction of inspired oxygen	FIO_2
Inspiratory reserve volume	IRV
Expiratory reserve volume	ERV
Residual volume	RV
Total lung capacity	TLC
Functional residual capacity	FRC
Vital capacity	VC
Forced expiratory volume in one second	FEV_1
Forced expiratory volume	FEV
Forced vital capacity	FEC
Peak expiratory flow rate	PEFR
Forced expiratory time	FET
Usual interstitial pneumonia	UIP
Desquamative interstitial pneumonia	DIP
Lymphocytic interstitial pneumonia	LIP
Pulmonary lymphoid hyperplasia	PLH
Nodular lymphoid hyperplasia	NLH
Idiopathic pulmonary fibrosis	IPF
Bronchiolitis obliterans	BO
Bronchiolitis obliterans with organizing pneumonia	BOOP
Histocompatibilty complex antigens	HLA
Transforming growth factor-B	TGF-B
Macrophage chemotatic protein-I	MCP-I
Interleukin-I	IL-I
Tumor necrosis factor alpha	TNF-α
Interleukin-2	IL-2
Intercellular leukocyte adhesion molecules	ICAM
Endothelial leukocyte adhesion molecules	ELAM
Broncho-alveolar lavage	BAL
Human T-cell lymphotropic virus type-1	HTIV-1
Open lung biopsy	OLB
Acquired immune-deficiency disease	AIDS
Polymerase chain reaction	PCR

CONTENTS

INTRODUCTION

INTRODUCTION & AIM OF THE ESSAY

Introduction :

The pulmonary interstitium provides the connective tissue structure that supports the lung. It extends in a continuous compartment from the mediastinum to the pleura.

The interstitial lung diseases (ILDs) are a heterogenous group of disorders that affects predominantly the lung parenchyma with minimal airway involvement. Detection and diagnosis, at times, are difficult. (Bernstein & Stulborg., 1994)

This heterogenous group of disorders follows a common pathogenic process. Interstitial lung disease involves recruitment of inflammatory cells into the pulmonary interstitium, alveolar walls, peri-alveolar tissues, this pathogenic process is referred to as " alveolitis " which lead to thickening of the alveolar walls. (Bokulic & Hilman.,1994)

The mechanism by which alveolitis and epithelial injury evolve into interstitial fibrosis are unknown. Progression of interstitial inflammation to fibrosis is because of factors other than just the presence of inflammatory cells. Some types of ILD suggests a role for immunologically mediated injury. (Marguerie., 1990)

There is no satisfactory clinical or pathological classification of these diseases, reflecting the poor understanding of both etiology and natural history. (Williams &Phelan.,1984)

Aim of the essay :

The essay will present the different aspects of interstitial lung diseases (ILDs) as regards etiology, pathology , clinical picture , diagnosis and management.

ANATOMY

ANATOMY OF THE RESPIRATORY SYSTEM

The respiratory system is divided into upper and lower airways. The upper airway includes the nose, paranasal sinuses and the pharynx, the lower airways includes the larynx, trachea and its division. The trachea divides into two main bronchi which in trun divide into lobar, then segmental and subsegmental bronchi, these divide into bronchioles. (Dinwiddie.,1990)

Each bronchial division is called a generation. The airways systemically branch over an average of [23] generations of dichotomous branching ending eventually in blind sac. The airways from the mouth through the trachea constitute the conducting airways.The last six to seven generation of these airways are connected to tightly packed alveoli, airway chambers in which gas exchange take place. (Moore.,1992)

Lungs are the essential organ of respiration and are attached to the heart and trachea by their roots and the pulmonary ligaments. It contains non respiratory tissues, which are nourished by the bronchial arteries and drained by pulmonary veins. (Chung.,1992)

Lungs are cone shaped and described as having an apex, base ,costal surface and medial surface. The apex is rounded and raises into the root of the neck about one inch above the middle thrid of the clavicle. The base is concave and is closely related to the thoracic surface of the diaphragm. The costal surface is convex and is closely related to costal cartilages, the ribs and the intercostal muscles. The medial surfaces are concave and are separated from each other by the mediastinum. The medial surface of each lung has a roughly triangular shaped area known as the hilum, through which the components of the root of the lung enter or leave the lung, these consist of bronchus,

a pulmonary artery, two pulmonary veins, a bronchial artery, bronchial veins, nerves and lymph vessels. (Wilson.,1981)

The right lung is divided into upper, middle and lower lobes by oblique and horizontal fissures. It has three lobers [secondary bronchi] and 10 segmental [tertiary] bronchi. The left lung has two lobes, upper and lower. It has an oblique fissure that follows the line of the sixth rib and has 2 lobar [secondary] bronchi. The broncho- pulmonary segment is the anatomical, functional and surgical unit of the lungs. (Chung., 1992)

Tracheobronchial tree :

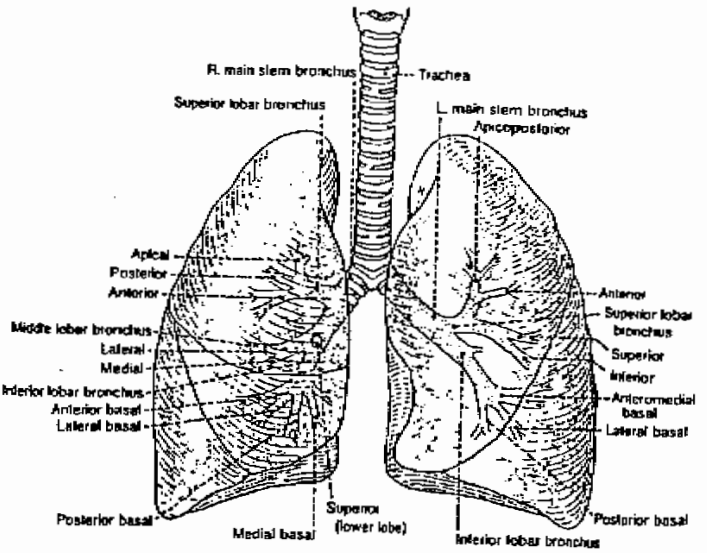
The lungs are supplied by a single common airway, the trachea, which passes down from the larynx into the thorax. It divides into the right and left main bronchi. The right main bronchus wider and shorter than the left, it follows the line of the trachea more closely than the left. The right main bronchus gives rise to three lober bronchi:

1- Right upper lobe bronchus; it arises from the lateral wall of the main bronchus at an angle of 90.
2-Middle lobe bronchus; it arises from the anterior wall of the bronchus.
3-Lower lobe bronchus; it is the downword continuation of the main bronchus .

The left main bronchus gives rise to two lober bronchi:

1-Upper lobe bronchus.
2-Lower lobe bronchus.

The lober bronchi each divide into two or more segmental bronch. The bronchi become progressively narrower and eventually become bronchioles. These divide further until the terminal bronchioles are reached; these lead to respiratory bronchioles. (Horse Field.,1981)



Fi.g (1): Anterior view of the trachea, bronchi and lungs *[From: Chung, 1992]*

Upto [50] respiratory bronchioles are arised from each terminal bronchioles. Respiratory bronchioles continue to branch into 2-5 further subdivision, until they end in alveolar ducts from which arise the alveolar sacs and then most of the alveoli themselves { alveoli are the final blind-ends of the respiratory tract }. Those parts of the respirtory tracts distal to the terminal bronchiole and concerned with gas exchange are the terminal respiratory units, reffered to collactively as lung parenchyma and sometimes individually as acini or primary lobes. There are aproximatly [300] million alveoli in the average adult lung, and each alveolus is about [250 um] in diameter. At brith, an infant has a full complement of airways but only about [25] million alveoli, thus though no new airways form after birth, most of the alveoli present in the adult have developed during childhood. (Cade & Pain.,1988)

The alveolar wall is composed of epithelial cells in close association with a capillary endothelial network. Approximately 95% of the alveolar surface is lined with squamous { type- I } pneumocytes with the remainder being granular {type- II} pneumocytes . (Mason ,et al.,1991)

Type-I pneumocytes are less abundant than type- II pneumocytes but cover more of the alveolar surface. (Crapo, et al., 1982)

Type-I cells serve two functions.In areas where their cytoplasm is thinnest, gas exchange takes place, oxygen diffuses out of the alveolus into the blood of the pulmonary capillaries, and carbon dioxide diffuses out of the blood into the alveolus.Alveolar air is separated from the blood by a very thin blood - air [exchange] barrier composed of alveolar epithelium, alveolar basal lamina, capillary basal lamina and capillary epithelium [endothelium]. In man areas of the lung, the basal lamina of the alveolus fuses with that of the capillary, so little or no space exists between epithelial layers. The

epithelial linings of the alveolus and capillary, together with the intervening connective tissue are referred to as the respiratory membrane.Where the type-I alveolar cells are thicker and there is more connective tissue, water and proteins are exchanged between the interstitial spaces and lymphatic capillaries. (Weinreb.,1984)

Type-I pneumocytes, thin, flat cells that are sensitive to noxious agents do not possess the ability to regenerate when injured.

Type-II pneumocytes are more resistant to injury than type-I pneumocytes and can proliferate alonge the alveolar surface and transform into type-1 pneumocytes. (Rennard, et al.,1983)

Type -II pneumocytes is a cuboidal secretory cell. It is the source of the surface active material, pulmonary surfactant, which coats the inner surface of the alveoli and lowers surface tension. Macrophages present through out the alveolar wall protect the lung as they phagocytase and lyse small foreign particles in inhaled air. (Weinreb.,1984)

Injury to type -II pneumocytes may lead to formation of lamellar bodies from lipid and protein substances within the cells.
(Katzenstein,et al.,1986)

The alterations in the production and excreation of surfactant- B can lead to alveolar collapse. (Burkardt.,1989)

Communictions between adjacent alveoli via pores of 10-15u in diameter permit gas flow between alveoli and segments of the lung {collateral ventilation}.These alveolar pores { pores of Kohn } may help prevent collapse when a local airway is occluded . Other alternative pathways for