RESTRICITVE LUNG DISEASE

- RLD are a category of respiratory disease characterized by a loss of lung compliance causing incomplete lung expansion and increase lung stiffness.
- In RLD both lung volumes and capacities are getting decreased.
- This occurs either because of :-

1) Alteration in lung parenchyma.
2) Disease of the pleura, chest wall or neuromuscular apparatus.

- Physiologically restrictive lung diseases.

Lung volumes & capacities	Effect of RLD
Tidal volume	Decreases
Inspiratory reserve volume	Decreases
Expiratory reserve volume	Decreases
Residual volume	Decreases/normal
Vital capacity	Decreases
Total lung capacity	Decreases
Functional residual capacity	Decreases
SPIROMETRY MEASURES	
FVC	Low
FEV_1	Decreases
FEV_1/FVC	Normal/high

- Restrictive lung diseases are divided into two group :-

1) Intrinsic lung diseases.(diseases of lung parenchyma)
2) Extrinsic lung diseases.(extra parenchymal diseases)

References: Cash text book of chest, heart and vascular disorders
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INTRINSIC LUNG DISEASES

- These diseases causes either :-

-Inflammation and/or scarring of lung tissue.(Interstitial lung disease)

OR

-Fill the air spaces with exudate and debris.(Pneumonitis)

PULMONARY CAUSES OF RLD :-

- Idiopathic Pulmonary Fibrosis
- Coal Worker's Pneumoconiosis
- Silicosis
- Asbestosis
- Pneumonia
- Pleural effusion
- Bronchogenic Carcinoma
- ARDS
- Sarcoidosis

EXTRINSIC LUNG DISEASE

- The chest wall, pleura, and respiratory muscles are the component of the respiratory pump.
- The disorder of these structures cause lung restriction and impair respiratory function.
- These are grouped as :

References: Cash text book of chest, heart and vascular disorders
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A)Neuromuscular diseases.

NEUROMUSCULAR CAUSES :-

- Spinal Cord Injury
- Amyotropic Lateral Sclerosis
- Poliomyelitis
- Gullian-Barre Syndrome
- Myasthenia Gravis
- Tetanus
- Duchenne's Muscular Dystrophy
- Other Muscular Dystrophies

B)Non-muscular diseases of the chest wall.

(1) Musculoskeletal cause: kyphosis & scoliosis, Diaphragmatic Paralysis or Paresis

Ankylosing spondylitis

3) NUTRITIONAL AND METABOLIC CAUSES :-

- Obesity

4) Cardiac causes

- Pulmonary embolism

5) CONNECTIVE TISSUE CAUSES :-

- Rheumatoid Arthritis
- Systemic Lupus Erythematosus

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- Scleroderma

- Polymyositis

- Dermatomyositis

) IMMUNOLOGIC CAUSES :-

- Goodpasture's Syndrome

- Wegener's Granulomatosis

7) REPRODUCTIVE CAUSES :-

- Pregnancy

8) TRAUMATIC CAUSES :-

- Crush Injuries

- Penetrating Wounds

- Thermal Trauma

9) THERAPEUTIC CAUSES :-

- Surgical Therapy

- Drug Therapy

- Radiation Therapy

PATHOGENESIS :

COMPLIANCE :-

- With RLD, chest wall or lung compliance or both decreases.

LUNG VOLUMES :-

- Generally all the lung volumes and capacities are getting decreased.

References: Cash text book of chest, heart and vascular disorders
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- Decrease in TLC & FRC are a direct result of a decrease in lung compliance

SIGNS & SYMPTOMS OF RLD:-

- SIGNS :

- Tachypnea

- Hypoxemia

- Decreased lung volume

- Decreased diffusion capacity

- Decreased breath sounds

- Pulmonary hypertension

- Clubbing

- Cynosis

- SYMPTOMS :

- Dyspnea

- Cough

- Weight loss

- Muscle wasting

PATHOPHYSIOLOGY

INTRINSIC LUNG DISEASES :

- Diffuse parenchymal disorders cause reduction in all lung volumes.

- This is produced by excessive elastic recoil of the lungs.

- Expiratory flows are reduced in proportion to lung volumes.

References: Cash text book of chest, heart and vascular disorders
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- Arterial hypoxemia is caused by ventilation/perfusion mismatch.
- Impaired diffusion of oxygen will cause exercise-induced desaturation.
- Hyperventilation at rest secondary

EXTRINSIC LUNG DISEASES :

- Diseases of the pleura, thoracic cage, decreases the compliance of respiratory system.
- There is reduction in lung volumes.
- Secondarily, atelectasis occurs leading to V/Q mismatch leading to hypoxemia.
- The thoracic cage and neuromuscular structures are a part of respiratory system.
- Any disease of these structures will cause restrictive disease and ventilatory dysfunction.

IDIOPATHIC PULMONARY FIBROSIS

SYNONYMS : cryptogenic fibrosing alveolitis, interstitial pneumonitis, and Hanman-Rich syndrome.

- It is an inflammatory process involving all the components of the alveolar wall that progresses to gross distortion of lung architecture.

PATHOPHYSIOLOGY

- Lung involvement in IPF shows patchy focal lesions scattered throughout both lungs.
- Followed by inflammatory changes and then scar and become fibrotic, distorting alveolar septa and the capillary network.

  Alveolar spaces become irregular in size and shape and there is significant progressive destruction of capillary bed

- Now these changes combine to cause,

  - decreased lung compliance

References: Cash text book of chest, heart and vascular disorders
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- decreased lung volumes

- increased V/Q mismatching

- decreased surface area for gas exchange

- decreased diffusion capacity

- increased pulmonary arterial pressure

- which increase the work of right ventricle

- increased work of breathing

- increased caloric requirement

- decreased functional capacity.

- Two major pathologic component of IPF are :

  1) inflamatory process in the alveolar wall.

  (sometimes called alveolitis)

  2) scaring or fibrotic process secondary to active inflammation.

- Both of this components occurs simultaneously within the lung.

<u>COAL WORKERS' PNEUMOCONIOSIS</u>

- caused by inhalation of coal dust. It is divided into simple CWP & complicated CWP
- It is caused by repeated inhalation of coal dust over a prolonged period of time i.e 10-12 yrs.
- Complicated CWP, sometimes called as progressive massive fibrosis occur only after longer exposure to coal dust.
- Antharcite coal is more hazardous than bituminous in the development of this disease.

References: Cash text book of chest, heart and vascular disorders
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PATHOPHYSIOLOGY

- Coal macules – focal collection of coal dust with little tissue reaction in terms of either cellular infilteration or fibrosis.
- These coal macules are located at the division of the respiratory bronchioles and are often associated with fibrotic emphysema.
- Lymph nodes are enlarged and homogeneously pigmented and are firm and not fibrotic.
- The pleural surface appear black owing to deposition of coal dust.
- Less than 5% cases progress to complicated CWP.
- The mechanism for the progression of simple to complicated CWP is unknown.
- It has been suggested simple CWP may progress, when it combines with infection, or silicosis, or tuberculosis or altered immunologic mechanism.
- Complicated CWP results in zones of dense fibrosis in the apical segments in one or both the lungs.
- These zones are made of dense, acellular, collagenous, black pigmented tissue.
- The normal lung parenchyma is completely replaced, and the blood vessels in that area show obliterative arteritis.
- These fibrous zone completely replace the entire upper lobe.

SILICOSIS

- It is one of the occupational pneumoconisosis, is a fibrotic lung disease caused by the inhalation of the inorganic dust known as free or crystalline silicon dioxide.

PATHOPHYSIOLOGY

- Inhaled silica causes macrophages to enter the area to ingest these particles.
- But cytotoxic effect of silica destroy these macrophages.

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

- This process release lysosomal enzymes, which induces the progressive formation of collagen, which eventually become fibrotic.

- An other characteristic of silicosis is formation of acellular nodules composed of connective tissue called silicotic nodules.
- Initially this nodules are small but as the disease progress it becomes large.
- Silicosis normally affect the upper lobes of the lung more than lower lobes.

PNEUMONIA

- It is a inflammatory process of the lung parenchyma.
- The disease may be classified anatomically as lobular, lobar, or segmental. Bilateral lobular pneumonia is termed bronchopneumonia.
- The commonest cause is infection by bacteria such as, streptococcus pneumoniae/pyogenes, staphylococcus pyogenes and klebsiella pneumoniae, mycoplasmal pneumoniae. Legionella pneumophilia causes pneumonia known as legionnaires disease.

PATHOPHYSIOLOGY

- The most common route for infection leading to pneumonia are inhalation and aspiration.
- When the causative organism is bacteria, the first response will be outpouring of edema fluid.
- This is followed rapidly by the appearance of polymorphonuclear leuckocytes that are involved in active phagocytosis of the bacteria, and than fibrin is depositied in the inflamed area.

References: Cash text book of chest, heart and vascular disorders
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- Clinically, bacterial pneumonia usually has an abrupt onset and is characterized by lobar consolidation, high fever, chills, dyspnea, tachypnea, productive cough, pleuritic pain, and leukocytosis.
- When the causative agent is viral, the virus first localizes in respiratory epithelial cells and causes destruction of the cilia and mucosal surface, leading to loss of mucociliary function.
- If viral infection reaches the level of alveoli, there may be edema, hemorrhage, hyaline membrane formation, and possibly the development of adult respiratory distress syndrome.
- Primary viral pneumonia is a serious disease with diffuse infiltrates, extensive parenchymal injury, and severe hypoxemia.
- Clinically, viral pneumonia usually has an insidious onset and is characterized by patchy diffuse bronchopulmonary infiltrates, moderate fever, dyspnea, tachypnea, nonproductive cough, myalgia, and normal WBC count.

Signs and symptoms

Initially symptoms are similar to that of a cold followed by:

- a high fever (pyrexia)
- chills
- a productive cough

Sputum may be discoloured and may become blood-stained as the pneumonia progresses. The following may also occur:

- dyspnoea
- sharp chest pain
- worsening cough
- headaches

References: Cash text book of chest, heart and vascular disorders
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- malaise
- muscle pains
- cyanosis due to poorly oxygenated blood
- loss of appetite
- rapid breathing
- wheezing or grunting during breathing
- intercostal muscle recession during breathing
- vomiting
- Physical examination
    - crackles and wheezing may be heard while auscultating

INVESTIGATIONS

- The X-ray will show decreased lung expansion and opacity on the affected side

- Sputum samples and blood tests
    - done to diagnose the type of pneumonia that is present
    - sputum test is done to determine whether it is a fungal or bacterial infection
    - blood test is done to examine the White Blood Cell count of the involved patient
    - this can be used to indicate the severity of the pneumonia, as well as to determine whether it is a viral or bacterial infection.
    - bacterial infection would result in a blood count that has an increased amount of neutrophils
    - a blood count that has an increased amount of lymphocytes would indicate a viral infection.

Physiotherapy management

 Modified postural drainage - this allows gravity to drain secretions from specific segments of the lungs

 Shaking and vibrations - to mobilize secretions

 Coughing and huffing exercises - to expectorate secretions

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

 Administer humidification - to mobilize secretions

 Breathing exercises - Localized and Diaphragmatic

 IPPB administration - to increase lung volumes

 Mobilization of the patient - done to increase air entry, increase chest expansion, and to loosen secretions

ADULT RESPIRATORY DISTRESS SYNDROME

- ARDS is a clinical syndrome caused by acute lung injury and characterized by severe hypoxemia and increased permeability of the alveolar capillary membrane.

PATHOPHYSIOLOGY

- Primary pathologic change is increase permeability of the microvascular pulmonary membrane.
- This will allow excess fluid and plasma protein to move out of vascular channel.
- This fluid leak into the interstitial tisssue and then crosses the alveolar epithelium to fill the alveoli.
- The change from air-filled to fluid-filled organ leads to-

  - decrease in compliance of the lung & all lung volumes and capacities.

  - increase in work of breathing.

  - pulmonary vascular resistance is increased and intrapulmonary right-to-left shunt takes place.

  - V/Q mismatching is increased

  - gas exchange is reduced.

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

- surfactant production is decreased.

- The significant atelectasis due to edema in the interstitial space leads to increase in pressure on the adjacent bronchioles and alveoli.
- Acute phase- ARDS resolve completely within few months.
- Sub-acute phase- as alveolar fibrosis and capillary obliteration develop within the lung, it leads to chronic restrictive lung dysfuntion.

Clinical manifestations

The signs and symptoms of ARDS are characterized by severe defects in oxygenation requiring the use of mechanial ventilation and pulmonary edema with no underlying elevated left atrial pressures that ould explain the presentaton Because ARDS is used as an umbrella term to describe pulmonary dysfunction after a direct trauma or immune disorder, the clinical manifestations vary between professionals.

When treating a patient with ARDS, it is important to keep in mind that there will be both a physiological and psychological compenent to take into account. Because people suffering from ARDS will experience a difficult time to catch their beath, they will become highly anxious and stressed, which could exacerbate the condition. This is where physiotherapists play a key role in limiting the individual's psychological distress by educating the patient in regards to what they are going through as well as trying to keep the patient as calm as possible. With regards to the physiological management of the condition, there are several interventions that can be used to limit the negative effects of ARDS, which can include: positioning of the patient, suctioning, and the rehabilitation of muscle strength once the patient has recovered from the condition and are now focusing on returning to their original state.

Positioning

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

According to many recent studies, the most effective position in treating ARDS is prone. The dependent lung is compressed when lying in supine, Based on reviewing a list of studies on ARDS treatment, a suggested procedure has been described by Hough (2014) to safely move the patient from a supine position to a prone position:

- Explain to the patient, with reassurance, that they will be safe and obtain consent if they are able to communicate.
- Close eyes and protect with gel or pad.
- Place the patient's palms against their thighs, thumbs upwards, elbows straight and shoulders neutral.
- Slide the patient to the edge using a gliding sheet.
- Roll patient into the lateral position using the underneath sheet.
- Roll patient into prone.
- 'Swimmers position' - elbow in which the head is semi-rotated should be flexed to no more than 90° to avoid ulnar nerve stretch, and the other arm internally rotated by the side.
- Ensure that women's breasts or men's genitals are not compressed.
- Place two pillows under each shin to prevent peroneal nerve stretch, positioning them to avoid knee and toe pressure from mattress.

Suctioning

Because the patient will have difficulty breathing, they will not be capable of performing any breathing techniques that are commonly utilized to excrete the secretions within their lungs. Because of this, suctioning will most often be used to clear out the secretions to ensure that no infections occur within the lung tissues.

Mechanical Ventilation

Because ARDS patients experience hypoxemia and a high work of breathing, many of them experience ventilatory failure with hypercapnia and respiratory acidosis[While working with a

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

patient suffering from ARDS, it is important to stabilize the breahting to ensure normal arterial blood oxygenation; this is achieved by raising the fraction of inspired oxygen (FIO2) and applying positive end-expiratory pressure (PEEP). Unfortunately, there is a lack of evidence that compares the effectiveness of the numerous mechanical ventilation techniques, therefore, we do not know which method to use in ARDS patients.

PLEURAL EFFUSION

- It is the accumulation of the fluid within the pleural space.
- The fluid is transudate if it has a low protein content and accumulates owing to change in hydrostatic pressure within the pleural capillaries.
- The fluid is exudate if it has high protein content and accumulates because of change in permeability of pleural surfaces.

PATHOPHYSIOLOGY

- The capillary in the parietal pleura receive blood via high pressure systemic arterial circulation.
- The capillary in the the visceral pleura receive blood via low pressure pulmonary circulation.
- Because of this pressure gradient fluid is constantly moving from parietal pleural capillaries to pleural space and is than reabsorbed into the visceral pleural capillaries.
- Approx 5-10 liters of fluid pass through pleural space each day using this route.
- Normally fluid formation and resorption are balanced and fluid donot accumulate in the pleural space.
- When this balanced is disrupted due to any cause it leads to accumulation of pleural fluid in the space and thus causes restriction in lung function by donot allowing the lungs to expand.

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

- Transudative pleural effusion are associated with an elevation in the hydrostatic pressure in the pleural capillaries.
- This is commonly due to left-sided heart failure, right-sided heart failure or both.
- Because of increase in hydrostatic pressure, more fluid is moved out of pleural capillaries and less fluid is reabsorbed.
- Therefore there is excess pleural fluid in pleural space, causing bilateral pleural effusions.
- Congestive heart failure is the most common cause of transudative pleural effusion.
- Exudative pleural effusions are associated with increase permeability of the pleural surfaces that allows protein and excess fluid to move into the pleural space.
- Therefore in exudative pleural effusion pleura is involved in pathologic process.
- Most commonly involved in inflammatory process or with neoplastic disease.
- Inflammatory process such as pneumonia, T.B, pulmonary emboli with infarction cause disruption of the normal pleural permeability.
- Cancer can also cause disruption of normal pleural permeability, either by direct extension of lung tumour in to pleural surface or indirectly tumour cells are spread via lymphatics.

PHYSIOTHERAPY MANAGEMENT

- Positioning for effective drainage
- Diaphragmatic breathing exercises
- Thoracic expansion exercises
- Exercises to mobilize the chest
- Secretion clearing strategies: :manual techniques to loosen secretions including oercussion, vibration and shaking
- Humidification
- Huffing/ coughing

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

- ACBT

PHYSIOTHERAPY MANAGEMENT OF PULMONARY FIBROSIS

The goals of physiotherapy management in Pulmonary fibrosis include the following:

1. Maximize the patient's quality of life, general health and wellbeing.
2. Educate the patient about PF, self management, ceasing of smoking, prevention of infections.
3. Optimize alveolar ventilation.
4. Optimize lung volumes and capacities.
5. Reduce the work of breathing.
6. Maximize aerobic capacity and efficiency of oxygen transport.
7. Optimize physical endurance and exercise capacity.
8. Optimize general muscle strength and thereby peripheral oxygen extraction.

Patient monitoring includes dyspnea, respiratory distress, breathing pattern(depth and frequency), arterial saturation, heart rate, blood pressure, and rate pressure product. patients with cardiac dysfunction or low arterial oxygen tensions require ECG monitoring, particularly during exercise. Breathlessness is accessed using a modified version of the Borg scale of perceived exertion.

The primary interventions for maximizing cardiopulmonary function and oxygen transport in patients with PF include some combination of education, aerobic exercise, strengthening exercises, chest wall mobility exercises, body positioning, coughing, relaxation techniques, pacing and energy and energy conservation. An ergonomic assessment of home and work environments may be indicated to maximize function in these settings.

Education includes information in preventative health practices, such as removing from the causative environment, flu shots, smoking reduction and cessation, weight control, hydration, relaxation, energy conservation.

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics

During aerobic exercise, patients with PF are prone to arterial desaturation. Patients who desaturate during sleep, require supplemental oxygen during exercise.

References: Cash text book of chest, heart and vascular disorders
Cardiorespiratory physiotherapy: Adults and pediatrics