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W3_L5_Clinical Diagnosis of Restrictive Lung Disease - Part 1

Good morning one and all. I am Dr. Vigneshwaran, Professor of medicine from Chettinad Academy of Research and Education. The topic we are going to discuss under PFT today is clinical diagnosis of restrictive lung disease. The objectives of my talk will be, first we will see what do we mean by the term restrictive lung disease. Then we will see what are the diseases that come under the category of restrictive lung disease.

Then we will see how to differentiate restrictive lung disease from obstructive lung disease. Then we will come to the topic per se which is how to clinically diagnose a case of restrictive lung disease and finally to understand it better we will have some clinical scenarios to understand it better. Now we will start with a brief introduction. Now respiratory diseases are grouped under three categories.

One is obstructive lung disease where the problem is in the airway. Second restrictive lung disease where the problem can be in the lung parenchyma or in the pleura or in the chest wall or in the respiratory muscles or the neuromuscular junction or the nerves which supply these respiratory muscles or in the respiratory center in the brain. The third type of respiratory disease is the pulmonary vascular disease where the problem is in the pulmonary vasculature. Now to understand this in the diagrammatic way, now we can see that the respiratory diseases come under three categories. So, first is the airway disease.

Here the obstructive airway disease, the problem is in the airway. Second is the restrictive lung disease where the problem can be in the lung parenchyma as in this case or it could be in the, so lung parenchyma disease or problem could be in the pleural cavity or in the thoracic cage or in the respiratory muscles or in the neuromuscular junction or in the nerves which supply these respiratory muscles or in the respiratory center of the brain. So, everything comes under the category of restrictive lung disease. The third disease of respiratory diseases will be pulmonary circulation diseases or pulmonary vascular diseases. Now we will go to the first objective of our talk.

What is the meaning of the term restrictive lung disease? Restrictive lung disease is a heterogeneous group of lung diseases. It is characterized by limitation and the ability of the lung to expand during inspiration. Now we will explain these terms better. What do we understand by the word heterogeneous? Heterogeneous means different types of lung

diseases come under this category and the commonality among all these different types of lung diseases is that there is a difficulty or limitation in the ability of the lung to expand during inspiration. What it means is, it means that there is decreased compliance of the lung or distensibility of the lung or elasticity of the lung.

In spirometry all these restrictive lung diseases have reduced total lung capacity and forced vital capacity. We can see this in this spirometry readings which shows the difference between normal and restrictive. What we can observe in this is that the total lung capacity which is usually around 6 liters which includes residual volume, expiratory reserve volume, tidal volume and inspiratory reserve volume. Here in restrictive lung disease the total lung capacity is reduced. That is reduction in almost all the types of volumes you see in spirometry.

There is also reduction in the forced vital capacity. Forced vital capacity is nothing but total lung capacity minus the residual volume. So, as you can see in restrictive lung diseases the total lung capacity as well as the forced vital capacity are reduced. Now this is also can be seen in a flow volume loop. We can see that this is the normal flow volume loop.

Now see what happens in restrictive lung disease is that it is the same contour as a normal lung but what happens is that the loop is small. So, this is the flow volume loop changes which occur in a restrictive lung disease. Coming to the usage of this total lung capacity, total lung capacity can be used to grade the severity of the restrictive lung disease. This grading system is given by American Thoracic Society and by that we can grade the severity of the restrictive lung disease. What it says is the total lung capacity has to be less than 80 percent to call it a restrictive lung disease and it can be graded into grade 1 which is where the total lung capacity is 70 to 80 percent predicted, grade 2 it is 60 to 70 percent predicted, grade 3 50 to 60 percent predicted and grade 4 which is severe restrictive lung disease where the total lung capacity is less than 50 percentage of predicted.

Now coming to the second objective what are the diseases that come under the category of restrictive lung disease? Now we already have told this that restrictive lung disease means a heterogeneous group of lung diseases. So, the restrictive lung disease the problem can be in the parenchyma of the lung. It is like a diffuse parenchyma lung disease or the problem can be in the flora which surrounds the lung or the problem can be in the chest wall or thoracic cage or it can be in the muscles which are there in the respiratory muscles or it can be in the neuromuscular junction between the nerves and the muscle or it can be in the nerves or it can be in the respiratory center in the brain. Now what are the diseases that come under these categories? If we start from below that is the lung parenchyma, we have it is otherwise called as diffuse parenchyma lung disease is otherwise called as interstitial lung disease or ILD. We have two important causes in that classification wise it is called ILD of known cause, ILD of unknown cause.

ILD of known cause is because of exposure to environment related allergens. It could be inorganic materials like asbestos, silicosis or it could be exposure to organic materials like which causes hypersensitivity pneumonitis. Second category is ILD of unknown cause where the most common disease is idiopathic pulmonary fibrosis. Now coming to the pleural problem, the pleural problem which cause restrictive lung disease could be pleural effusion or pneumothorax. Then coming to thoracic cage, the problems could be congenital or acquired conditions in the thoracic cage.

Congenital could be a sternal deformity or a spinal deformity. Sternal deformity example being pectus carinatum, pectus excavatum. This spinal deformity namely kyphosis and scoliosis. Acquired disorders include ankylosing spondylitis and even a morbid obesity will restrict the chest wall from expanding. Then there is a muscle problem like muscle myopathies, muscular dystrophies, neuromuscular junction problem like the myasthenia gravis.

Nerve problems, when you consider the nerves which supply the respiratory muscles, we can divide into actually into three categories. Now consider this as the respiratory muscle. It is supplied by a nerve; it is supplied by a nerve which arises from the anterior horn cell. This is called lower motor neuron and this is controlled with the brain which is called the upper motor neuron. So, you have got three categories in this, an upper motor neuron up to the anterior horn cell.

So, then you have got a lower motor neuron starting in the anterior horn cell and the lower motor neuron the radical. So, you can have diseases here like the AM myotrophic lateral sclerosis where there is a problem which affects the upper motor neuron as well as lower motor neuron. Then a disease in the anterior horn cell which is a part of lower motor neuron, the classical disease being poliomyelitis and spinal muscular atrophy. Then you have got a disease because of involvement of the radical or the nerve. Here the classical disease is Guillain-Barre syndrome and then you can have problem with the respiratory center in the brain like because of some cerebral trauma or because of some drugs like opiates.

So, these are the diseases which come under the category of restrictive lung disease. Now we will just see it in a slightly more detail about what are the diseases which come under this restrictive lung disease. First, we will start with some diseases in the lung parenchyma which is also called as diffuse parenchyma lung disease also called as interstitial lung disease. Now first to understand that we should understand what is the meaning of the word interstitium. Now lung as such it starts with the airway, the airway divides that is the trachea divides into two bronchi, the right and the left bronchus and each of these divides into primary, secondary, tertiary bronchus and they again divide like that 22 generations and finally the 23rd generation only you get what is called as alveoli.

Now you know the importance of the lung, the lung is where there is the gas exchange. So, the alveoli and the surrounding blood vessels that is where the gas exchange occurs. So, what is interstitium? Interstitium is the space which is in between the alveoli and the blood vessel. So, this is where the gas exchange should occur. So let us consider there is an interstitial fibrosis that is thickening here, gas exchange becomes a problem.

That is the meaning of the importance of the word interstitium. Now we will see the diseases that come under interstitial lung disease. It is broadly classified into two, ILD of known cause and ILD of unknown cause. In ILD of known cause it can be due to exposure to some inorganic environmental pollutants like asbestosis and silicoses or it could be secondary to exposure to radiation or long-term exposure to some medications like methotrexate, amiodarone, etc. or it can be related to connective tissue disorders.

ILD can occur secondary to connective tissue disorders like rheumatoid arthritis and things. It can be secondary to some vasculitis, small vessel vasculitis like vaginis granulomatosis or a Christol syndrome or it can also be related to exposure to organic solvents. So, these all come under ILD of known cause. So, as I was telling about exposure to environmental pollutants, the size of the particular matter decides on where they get deposited. Let us consider the particulate matter size in micrometer is around 10.

It deposits in the tracheobronchial tree, airway, not in the alveoli or the interstitium. So, those are the ones which are prone for obstructive lung disease. Whereas with regards to asbestosis and silicoses, what happens is that their size is between 2.5 to 10 micrometer and they get deposited in the interstitium of the lung and that is why they are the reasons for interstitial lung disease. So, we have to know the importance of the particulate matters.

Then with regard to connective tissues, secondary to connective tissues, the whole spectrum can affect the interstitium of the lung because connective tissue disease means it involves the connective tissue. Connective tissue is the space, the space in between alveoli and the pulmonary vessels are also interstitium. As a result, so they are also prone for interstitial lung disease. So, the classical diseases being rheumatoid arthritis, systemic sclerosis or scleroderma, lupus erythematosus, polymyositis, demodermisitis, all can be associated with interstitial lung disease. Now with regards to the vasculitis, small vessel vasculitis which can cause interstitial lung disease, the two main diseases which come under them are vaginis granulomatosis which is also nowadays called as granulomatous angitis, is called as granulomatous disease with vasculitis and Christol syndrome.

Then we will now come to ILD of unknown cause. So, what are the ILD which we do not still know the real cause. The most important disease which come under this is called idiopathic interstitial pneumonia. So, in that the most common disease is idiopathic pulmonary fibrosis also called as IPF. And there are also other diseases like NSIP, non-

specific interstitial pneumonia, DIP, discriminatory interstitial pneumonia, CUP or cryptogenic organizing pneumonia.

These are also there. Now what you have to know about idiopathic pulmonary fibrosis is that they occur in smokers and it is usually elderly male above the fifth or sixth decade and the classical thing is they have got honeycombing appearance in the HRCT especially in the base of the lung. Sub plural honeycombing is what is the classical description of idiopathic pulmonary fibrosis. Now we will come to the diseases which come under pleural cavity. The two main diseases that come under pleural diseases will be pleural effusion and second will be pneumothorax. Pleural effusion as you all know is collection of fluid in the pleural cavity.

It is a collection of fluid. See what happens because of that the lung cannot expand, the lung is collapsed, the lung is collapsed lung cannot expand. Pneumothorax is air in the pleural cavity and here also what happens the lung is collapsed and it cannot expand. Now coming to diseases which come under chest wall diseases, chest wall diseases the diseases that come under this will be we divided them into congenital and acquired. In congenital we have got it could be a sternal deformity problem or a spinal deformity problem. Sternum is in the center, spinal is in the back.

Sternal deformity problems could be pectus carinatum where it is a pigeon chest. See here also you can see the lung there is a whole mediastinum is altered as a result expansion of the lung is difficult. And is pectus excavatum where the sternum is dipped inside because of which the lung cannot expand properly. The spinal problems could be gross scoliosis. See in an x-ray how much the, because of the scoliosis how much the chest is altered and here the lung cannot expand properly.

These are examples of congenital causes of chest wall diseases which cause restrictive lung disease. Now we will come to acquired causes. The two main causes here will be ankylosing spondylitis and second is morbid obesity. Now what do you know, what do you have to understand about ankylosing spondylitis is ankylosing spondylitis is a disease where there is inflammation of the intervertebral joint space. So what happens after some time because of this inflammation when they fuse it becomes rigid spine like it is called a bamboo spine and you can see the difference between this spine which is flexible and this spine which is not at all flexible fixed.

As a result, the lung chest cannot expand during inspiration. The other acquired cause is the morbid obesity. See because of morbid obesity lot of adipose tissue gets deposited here because of that the chest cannot expand. So, these are some chest wall causes of restrictive lung disease.

Now coming to the third neuromuscular. Neuromuscular means he has to involve the muscle or the respiratory muscles or the neuromuscular junction could be neuromuscular

junction problem or the nerve problem or the respiratory center problem. Now we will see what are the diseases that come under these categories. So, for that first we have to know what are the primary muscles of respiration. The two primary muscles of respiration are diaphragm and the intercostal muscles. Now in a disease what are the diseases that come under this will be.

Now let us first we start with the muscle disease. As we see we saw know that the respiratory muscle, the muscle disease, the muscle disease which causes respiratory which causes restrictive lung disease will be is the myopathies, muscular dystrophy especially Duchenne's muscular dystrophy, Baker's muscular dystrophy. Then coming to the neuromuscular junction, the classical neuromuscular junction problem being myasthenia gravis. Few words you have to know about myasthenia gravis. Myasthenia gravis is an autoimmune problem and what happens is at the neuromuscular junction conduction of the impulse to the muscle occurs through acetylcholine at the nerve terminals and they have got receptors in the muscle site.

So, acetylcholine will be secreted from the nerve site. You will have receptors in the muscle site. Now what happens in myasthenia gravis is antibodies are produced against these acetylcholine receptors. So, they go and block these acetylcholine receptors preventing acetylcholine from activating the muscles.

So, this is about myasthenia gravis. Now we will come to the third part that is we have finished muscles, respiratory muscles, in neuromuscular junction. Now we will concentrate on the nerves. Now as we discussed earlier, nerve problem could be at the, that is at the peripheral nerve anterior horn cell or the going up that is the upper motor neuron or the corticospinal tract. So accordingly, we have got three important problems here. One is amyotrophic lateral sclerosis where I already described that it is a disease where both the upper motor neuron and lower motor neuron are affected and you know the famous personality who was crippled by this disease namely the, namely Stephen Hawkins.

Now the second part of this, second disease which come under the neuromuscular disease will be where the problem is in the anterior horn cell, the classical disease being polio. You know about polio, it is a communicable disease and what happens is that it affects the virus, enterovirus affects the anterior horn cell and we know that because of good immunization it is almost nearing eradication. Now third disease is in the peripheral nerve that is the radical, so it is called Guillain-Barre syndrome also called as GBS where its antibodies are produced against myelin or axon and that gets destroyed because of its, there is a rapid ascending paralysis. So, these are the diseases that come under neuromuscular diseases. Now coming to the third topic, so we have completed what do you mean by the term restrictive lung disease, then we have seen what are the diseases that come under restrictive lung disease.

Third now we are going to see how to differentiate restrictive lung disease from obstructive lung disease. We have got a table here which demonstrates the differences very clearly. In obstructive lung disease the anatomy involved is the airway. So, we saw the trachea dividing into bronchus, bronchioles and I told you it goes through 22 generations and till around 16 to 17 generation it is only the airway.

So there the problem is in the airway. In restrictive lung disease the problem is in the lung parenchyma or it can be in the thorax or chest wall. The breathing difficulty which is experienced in an obstructive lung disease is during expiration. They cannot breathe out the air because of obstruction. Whereas we know in restrictive lung disease the problem is in during inspiration. The pathophysiology is that in obstructive lung disease the problem is increased airway resistance.

The pathophysiology in restrictive lung disease is decreased compliance of the lung or the thorax and in the pulmonary function test the most important thing which we observe in obstructive lung disease is decrease in the forced expiratory volume at first second. So, FEV1 is restricted and we can see that FEV1 by FEC will be less than 80 percentage of predicted that is what happens in obstructive lung disease. In restrictive lung disease the main thing is the total lung capacity and forced vital capacity are reduced. Now we will see that in a spirometry difference as we can see here in the restrictive lung disease, we already know the total this is the normal one. Here restrictive lung disease we saw that the total lung capacity and vital capacity both are reduced.

In obstructive lung disease because there is obstruction during expiration the air gets accumulated so what happens the residual volume increases so that which remains in the lung increases and as a result everything else increases so you get an increase in the total lung capacity or a forced vital capacity. Now the differences which we can observe in a flow volume loop will be in a obstructive lung disease there are three important things which are very useful to find it out. One is that the peak expiratory flow is not to this predicted value it is low. Second is that it is the even during expiration it has got a scooped-out appearance. So, this is a very classical thing this is a normal curve this is the scooped-out appearance in obstructive lung disease.

Third is it does not reach the 0 degree 0 so it is called an open loop. Now the flow volume loop with regards to restrictive lung disease which we have already seen is it is following the same contour but what is there is everything is small so it is a normal contour but a small loop. So, these are the differences between restrictive lung disease and obstructive lung disease.