Course Name: Pulmonary Function Test - Interpretation and Application in clinical practice
Professor Name: Dr Vigneshwaran
Department Name: Medicine
Institute Name: Chettinad Hospital and Research Institute
Week – 03
Lecture - 06

W3_L6_Clinical Diagnosis of Restrictive Lung Disease - Part 2

Now, coming to the main topic, how to clinically diagnose a case of restrictive lung disease? Now clinically diagnosing case means first symptom analysis or history taking, second next will be general examination, then will be examination of the system. This is how we clinically diagnose a restrictive lung disease. First, we will see what are the symptoms that help us to differentiate or find out that it is restrictive lung disease and find out the etiology. Now there are some cardinal symptoms which we ask in a case of restrictive lung disease. The six symptoms are cough, sputum production, hemoptysis, dyspnea, chest pain and wheeze. With the six symptoms only, we classify the lung diseases as we can use using this, we can identify whether it is an obstructive or a restrictive or the problem is somewhere else.

So, that can be found out with this three with the six cardinal symptoms and point about what is the meaning of the word hemoptysis? Hemoptysis is presence of blood in the sputum. Now among of these six cardinal symptoms to diagnose restrictive lung disease we have got certain symptoms which are important and these symptoms vary according to what is the type of the restrictive lung disease. Like we have got a diffuse parenchymal lung disease or ILD your symptoms will be different. Large chest wall disease it will be different, neuromuscular disease it will be different.

So, we will see what are these symptoms. The symptoms in diffuse parenchymal lung disease. The most important symptom in diffuse parenchymal lung disease is dyspnea, exertional dyspnea and it is a gradually progressive dyspnea. So, that is a classical symptom of a ILD or a diffuse parenchymal lung disease. Second is cough.

The cough is chronic cough but it is always dry the problem is in the interstitium. So, that is the second thing. Third they also have because there is so much dyspnea and all there is also their lung capacity is less because of that there is EC fatigue ability that is another important symptom of diffuse parenchymal lung disease. Now we know that diffuse parenchymal ILD can be occur because of several causes. If you ask carefully the history, we can find out what is the cause which what is the etiology of this interstitial lung disease.

First, we ask for history of asbestos exposure for more than 10 years. So, what do you do? We ask whether they are they have worked in factories which produces asbestos or they live in houses which are roofed by asbestos or in shipyards where an insulation material asbestos is used or we ask for history of exposure to silica like whether they are they blast with machines the concretes or they dispose of rubbish of concrete or mines which produces silica. So, these help us to identify that the probable ILD is due to silicosis. So, all both asbestos and silicosis require a history of exposure for more than 10 years. Now with regards to ILD secondary to radiation or drug exposure just ask the history about whether there was any radiotherapy for any chest problems like breast cancer or lymphomas.

So, and chronic intake of drugs which produce ILD like amethotrexate or amiodarone or nitrofuranetone. So, these drugs can cause interstitial lung disease. Now then ILD can we know can also be secondary to connective tissue disorders. So, we ask history for rheumatoid arthritis like whether there is any history of symmetrical polyarthritis for more than 3 months. So, interstitial polyarthritis is very important for rheumatoid arthritis.

Then whether there is any hand joint involvement, whether there is any early morning stiffness or more than one hour, whether there are any joint deformities. So, we know rheumatoid arthritis is known to produce it is erosive arthritis. So, it produces deformities like the swan neck or the bottom as deformity, whether there are any sicker symptoms which is dry mouth or dry eyes in the dry eyes, whether there is a family history of rheumatoid arthritis. So, these help us to identify whether it is ILD secondary to rheumatoid arthritis. And if it is related to scleroderma, we ask whether there is any joint stiffness, they cannot bend it properly, there is stiffness or there is because of skin thickening in scleroderma, they cannot open the mouth well.

So, difficulty in mouth opening and whether there is any obvious skin thickening we ask and we also ask for an important history called as Reynolds phenomenon. What is Reynolds phenomenon? Reynolds phenomenon is that is the distal vessels go for a vasospasm, arteries go for a vasospasm or exposure to cold. So, because of exposure to cold and vasospasm, what happens is that as you can see in that diagram, there is failure, that is no blood supply there. Because of that there is pain and numbness also. Now if you leave it like this for some time, what happens there is bluish discoloration and sometimes it can even go for gangrene, that is it is death of that tissue can occur.

So, these are the history you ask for a connective tissue disorder. Now with regards to ILD secondary to small vessel vasculitis, we saw two things, vaginis and chrystol syndrome. Here we ask vaginis granulomatosis which is also called as granulomatosis with polyangiitis, there is bloody discharge, sinusitis with bloody nasal discharge, cough with hemoptysis and presence of some palpable purpuras, vesicles and ulcers. So, we ask

for this history, whether they are present or not. With regards to chrystol syndome, we ask for history of allergic rhinosinusitis and is there any episode of asthmatic attacks.

So, this is the history for all these known causes. Now for ILD of unknown cause, it is found that idiopathic pulmonary fibrosis is the common and there is a strong association with smoking and as I told you it occurs in 56 decades of life, mostly in males and there is a strong family history also. And second similar idiopathic interstitial pneumonia is BISK primitive interstitial pneumonia, BIP. It also occurs commonly in active heavy smokers. So, this is the history we ask for a diffuse parenchymal lung disease or ILD.

Now what history is you can ask for a pleural and a chest wall disease. Now here the dyspnea, here will be subacute onset of dyspnea and gradually progressive that usually occurs with pleural effusion, but with regards pneumothorax, it will be acute and very severe and sudden. It will be, they suddenly become breathless. That is the feature of, that is the classical symptoms with dyspnea in pleural diseases. Now they can have cough, but it is usually non-productive or a dry cough.

The classical symptom with pleural disease is they can have a pleuritic type of chest pain. Now certain things, points about pleuritic chest pain are pleuritic chest pain is a chest pain which usually occurs in the lateral aspect of the chest. While lateral aspect it is where the parietal and visceral pleur are closer together and when they, even they are non-inflamed, there is no rubbing, but when they are inflamed, they are closer together and they rub. When that rub is there, they get severe pain. And the pain is, the classical description is a stabbing type of chest pain localized to the lateral aspect of the chest and worsens with deep inspiration and coughing.

When you take a deep inspiration, these rub each other. And among the several causes of chest pain, you can see that the pleurisy, the problem is in the lateral aspect. Lateral aspect is where the chest pain is. Now what are the symptoms in a neuromuscular disease? So neuromuscular disease, as I told you, the muscles, respiratory muscle problem, neuromuscular junction problem and nerve problem. What are the symptoms there? They also have dyspnea.

It can be gradual onset, subacute onset and gradually progressive like in a myasthenia gravis or rapid worsening as in a Guillain-Barre syndrome or in a myasthenia gravis, that is myasthenia gravis where suddenly it becomes worsened. They generally do not have any cough, sputum production, hemoptysis, chest pain or wheeze. Now how to identify the various types of neuromuscular diseases? That is only based on presence of the neurological symptoms which can first arrive at the diagnosis. Now to talk about a few neurological symptoms, in a myopathy, the respiratory muscles, the common myopathies being ditching muscular dystrophy and vagus muscular dystrophy, the weakness is predominantly proximal muscles. They have limb weakness, proximal muscles, leg

muscles, the proximal muscles are one which is weak and as a result they give history that they have difficulty in squirting past her.

They also have a definitive pattern of progression like what happens is at 2 years they have very minimal symptoms. At around 8 years what happens is there is weakness is starting to appear and they cannot get up from squirting posture and by 15 years they are crippled, they are wheelchair bound and other important thing is it is an extremely recessive disease. There is a family history of this disease. So, with this history will help us to identify whether it is probably a muscle disease. Now with regards to neuromuscular junction problem, ptosis is an important symptom.

Ptosis is drooping of the eyelids. You can see that ptosis, limb weakness, pharyngeal muscle weakness where there is difficulty in swallowing, respiratory muscle weakness, all these have diurnal variation in symptoms. What is the meaning? Morning they are better, evening as they go down, as the day progresses what happens is the weakness is becoming more. So, this is very obvious with regards to ptosis. You can see that mornings it is the lids are wide open, here evening it there is a drooping of the eyelids.

Now third among the neuromuscular disease is the nerve problem. The diseases we know are amyotrophic lateral sclerosis, then polio diseases and Guillain-Barre syndrome. What is important about amyotrophic lateral sclerosis? Amyotrophic lateral sclerosis, the problem is both in upper motor neuron and lower motor neuron, there is a progressive weakness. Initially it is asymmetric, only one area will be there, but then it becomes progressive and symmetric. In addition to weakness there is muscle wasting.

Usually, muscle wasting is slightly out of proportion to the weakness. The muscle wasting is quite gross. There is also fasciculation. This helps us to say it is slightly ALS. Then with regards to Guillain-Barre syndrome, the classical thing of Guillain-Barre syndrome is that the paralysis will be ascending.

It will start from the legs and the paralysis will go up and involve the upper limbs, then the facial muscles, then swelling difficulty, then the respiratory muscles. And the progression is quite rapid and it is usually preceded by a respiratory tract or an intestinal infection, just one week before, one to two weeks before this particular episode they can have this respiratory tract infection. Now this is with regards to history. Now coming to general examination of what features you will have in general examination of these patients with restrictive lung disease. Generally, there is no pallor or ictus, but you can have cyanosis.

You know cyanosis, bluish discoloration of the skin and the mucous membrane due to increase in the reduced hemoglobin. We know the parenchymal restrictive lung disease. There is a problem with the gas exchange and because of that the reduced hemoglobin

can increase and sinuses the manifestation of advanced restrictive lung disease. Then you can also have clubbing. It is also a sign of advanced lung disease.

Now clubbing you know is what? There is obliteration of the nail, nail bed angle. Because of that there is a bulbous enlargement of the distal phalanx and that is also an indicator of advanced lung disease. There is generally no lymphadenopathy and they can have pedal edema. Pedal edema is a marker in restrictive lung disease. It marks the development of car pulmonale.

What is the meaning of the word car pulmonale? car pulmonale is a right heart failure occurring secondary to a lung problem because of which what happens? Right heart becomes dilated and this occurs because of development of pulmonary artery hypertension. So, these are the general examination features in a case of a restrictive lung disease. Pedal signs generally the pulse and BP are normal but in the Guillain-Barre syndrome where there can be involvement of the autonomic nervous system the pulse and BP can have gross fluctuations.

This is called dysautonomia. Then respiratory rate is usually on the higher side there will be tachypneic more than 20 and a difference between tachypnea and hypopnea is tachypnea is shallow breathing and hypopnea is deep breathing. Otherwise, temperature is normal. These are the vital signs with regards to restrictive lung disease. Now coming to the examination of the respiratory system proper to identify these restrictive lung diseases. Now any the examination of a system involves inspection, palpation, percussion and auscultation.

First, we will see what are the findings in diffuse parenchymal lung disease or interstitial lung disease. Now in an inspection first we see the shape of the chest then we see the symmetry of the chest. The shape of the chest is usually elliptical as normal for us also it is elliptical. In obstructive lung disease only, it becomes barrel shape. The shape and symmetry are usually normal in a diffuse parenchymal lung disease.

Position of the trachea is also normal. Usually, it is in the midline or slightly to the right which is normal. Then there is no sternal or a spinal deformity. There are no abnormal pulsations but they can have dilated veins like elevated JVP in case of a car pulmonale. These are the inspiratory findings. Palpatory findings, shape and symmetry are normal.

Function of the trachea we know is confirmed. Then we see the movement of the chest. This is a very important point. Movement of the chest we do it in two ways. One the whole chest movements which is after a forced expiration the maximum inhalation is done. This expansion is difference is calculated and it is usually 3 to 5 centimeters.

Here it is reduced. So, in restrictive lung disease this chest expansion is reduced. We also can see the chest expansion in different areas. Generally, it is usually normal.

Percussion wise, percussion we do with the pleximeter and plexor and usually it is normal lung resonance with the diffuse parenchymal lung disease. Auscultation is very important to diagnose a diffuse parenchymal lung disease.

The breath sounds are usually normal acicular but the added sounds help us to diagnose. It is the diagnostic feature of the diffuse parenchymal lung disease also called as interstitial lung disease. What we see is we get end inspiratory fine crackles or rags. This is called as the Velcro crackles. What do you mean by the Velcro? It is like when you open a tight Velcro attachment.

So, what sound you get that is what is called as Velcro crepitations and vocal resonance is normal. So, these are all the examination findings of a diffuse parenchymal lung disease. Now we will see what will be the findings in a pleural and a chest wall disease. Now with here inspection wise, it is usually inspection wise the shape of the chest is normal. Symmetry in areas of pleural effusion or pneumothorax you can find that there is some amount of fullness.

Position of the trachea, this is important. In a pleural effusion and pneumothorax, the trachea is pushed to the opposite side and we can have sternal, these are chest wall diseases, pleural and chest wall diseases. You can have sternal deformities like a pectus carinatum where it is Pigeon chest or pectus excavatum where it is the sternum goes inside. Then you can have a spinal deformity, the two important types being kyphosis which is forward bending of the spine and scoliosis which is lateral bending of the spine. Then there are no abnormal positions and there are generally no dilated veins. Inspection wise, we can say the same shape and just say or symmetry are normal.

Position of the trachea as I told you it shifted to the opposite side as you can see in that image. Say right side at pleural effusion or pneumothorax the trachea shifted to the left side. Then movement of the chest, chest expansion is going to be reduced. But another important point to identify where is the side of the problem like a pleural effusion, the chest expansion we do it in different areas. So, we can see that in the infrascapular, infraxillary areas, interscapular areas the chest expansion is decreased on the side of the pleural effusion.

Percussion is very important to identify pleural diseases. You get a stony dull note to percussion. Normally when you percuss the lung you have got a normal lung resonance. In a pleural effusion it is stony dull note whereas in a pneumothorax it is hyper-resonant.

Percussion is very useful to find out pleural diseases. Percussion wise breath sounds it is of decreased intensity or sometimes it is even absent. Generally, there are no added sounds but in early cases like pleurisy you can hear the pleural rub which is actually a friction rub which increases with deep inspiration and coughing. It is also increased by pressing the steth on the chest. So, this occurs as we saw the inflamed pleura rubbing

against each other in a pleural disease. Pleural resonance is also decreased in areas of pleural effusion and pneumothorax.

Now coming to the third one that is examination findings in a neuromuscular disease. So, here we are going to discuss about neuromuscular that is respiratory muscles, neuromuscular junction, nerve problems. What will be the findings? Generally, inspection, palpation, percussion and auscultation of the respiratory system is usually normal. Like they can have they will have what they can have a single breath count is less otherwise the respiratory system examination is normal. How to diagnose then additional findings only neurological examination only will pick up the findings.

Now what are the findings which can be there in a muscle disease, muscular dystrophy they have this classical sign called Gower's sign. What is that is they climb there because there is proximal muscle weakness, they have to climb on their thighs to get up. So, this is very important to diagnose this muscular dystrophy and there is we know history wise there is a family history. Myasthenia gravis as we saw you can you can demonstrate ptosis of the eyes when they are asked to stare for a long time.

Slowly there is drooping of the eyelids. Then the problems in the nerves, amyotrophic lateral sclerosis it is identified by the gross muscle wasting. So, here you can see the thenar and hypothenar eminence this is the thenar, this is the hypothenar both are wasted and there is in addition tongue wasting as well as fasciculation. Fasciculations are very important to pick up the amyotrophic lateral sclerosis motor neuron disease. In polio you can see that they have got flaccid limbs and you know it is high sometimes hyper extending and it is a thin limb that is how folio is identified and Guillain-Barre syndrome where the classical thing is the ascending paralysis from legs to the upper limbs and the facial muscles. Now with these clinical scenarios we will understand this restrictive lung disease slightly in better way.

Now first clinical scenario, now a 60-year-old male who is a chronic smoker who smokes around 20 who has smoked around 20 packers. He has presented with intermittent cough. What do you mean by packers? So, one pack of cigarette per day per year is one pack year that is 20 cigarettes per day which is one pack for one year is one pack year. So, 20 pack years he has been smoking and he has developed intermittent cough, sputum production which is of mucoid type and all this has occurred over the past these symptoms are worsening over the or are present on almost all the days over in the past five years. In addition, whenever he develops some infection respiratory infection the sputum becomes yellowish and he starts to become dyspneic and there is also wheeze.

So, these with this history also we can make a diagnosis because there is wheeze and there is exposure to chronic smoking. Examination wise the patient is cyanosed you can see the lips being cyanosed central cyanosis and he has got first lip breathing he breathes

with the first lip. So, on exhalation the problem is there is a difficulty and he has got a barrel shaped chest. On auscultation there is scattered crepus and wheeze. Now what will be the diagnosis? Is it a restrictive lung disease? No, it is a classical example of an obstructive lung disease the problem is COPD.

So, this is the classical symptoms of obstructive lung disease. Now we will see this next scenario. Now a 50-year-old male laborer in construction industry he has been working for around 30 years. He drills concrete walls and floors with the drill machines in the daytime. In the evening, he clears all this rubbish this concrete rubbish in the evenings.

He never smokes or consumes alcohol. For the past one year he feels breathless while lifting heavy weights. He also feels tired and has to take rest in between work. He has been complaining of dry cough for the past six months. So, there is some environmental exposure in this case and he has had exertional dyspnea which is gradually progressive over the past one year. On examination there is tachypnea, respiratory rate is 30, there is clubbing, pan digital clubbing.

On auscultation he has got the most important thing is he has got some added sounds in the form of Velcro crackles present in both the infra-scapular and infra-axillary area. This is the chest x-ray which shows reticular opacities mostly in the upper lung. So, this is the classical disease, this is the classical presentation of a diffuse parenchymal lung disease causing a restrictive lung disease. So, this is a classical restrictive lung disease which is because of diffuse parenchymal lung disease and here it is secondary to environmental exposure to silica, silicosis, secondary to silicosis.

Now third clinical scenario. A forty-five-year-old female has history of multiple joint pains, symmetrical type of polyarthritis for the past ten years. There is also hand joint involvement, early morning stiffness for more than one hour and patient is on long term medications with methotrexate and leflonamide. These are the areas of pain and for the past six months initially she never had any dyspnea, for the past six months there has been breathlessness, dry cough and easy fatigue ability. On examination she is dyspneic, tachypneic, there is cyanosis, she has got finger deformities. S,o this is we saw already, this is the swan-neck deformity which is present and on RS examination what is there is the chest shape, tracheal position, percussion are all normal, auscultation wise there is fine end inspiratory crackles and this is heard in the bilateral infrascapular and interscapular areas.

So, this is again the Velcro crackles and this means it is a restrictive lung disease and here the restrictive lung disease is due to a lung problem that is diffuse for lung disease and it is secondary to a connective disorder like rheumatoid arthritis. Now coming to the next scenario, forty-five-year-old male with chest pain for the past two months. It is the chest pain is localized to the left infraxillary area and left infrascapular area. It is a sharp

stabbing pain increasing with deep inspiration. This is the site of the pain and there is dyspnea which is exertional for the past one month.

Dyspnea also increases if the patient is lying to the right side and he has got occasional cough and fever. In addition, he has lost around 4 kg of weight over the past two months. There is a history that his wife was treated for pulmonary tuberculosis cup two years before and was declared cured. On examination, patient looks ill-nourished, respiratory rate is high.

Inspection shows fullness in the left infraxillary area, fullness. Palpation wise there is decrease in the chest movements in this left infrascapular and infrascapular area. Percussion wise there is a stony dull note to percussion in the left infrascapular and infrascapular areas. So, stony dull note is very important to pick up plural diseases. Auscultation wise air entry is decreased in the left infrascapular area.

So, what is the diagnosis? This is also a restrictive lung disease. The problem here is not in the parenchyma of the lung. The problem is in the pleura. What is the problem in the pleura? The pleural problem is pleural effusion on the left side. So, this is another type of restrictive lung disease.

Now the last scenario to explain this. Now a 47-year-old male who has been complaining of pain over the neck, lower back and gluteal area for three days. Then within two days later he developed numbness of the palms and the soles. Then yesterday he developed difficulty in standing on toes.

He could not stand on the tip of the toes. On the, so he got himself admitted. On the day of admission, patient had difficulty in standing up from squatting posture or climbing stairs. In the subsequent two days, he could not lift his leg against gravity. Then he had difficulty in closure of the eyes and slurring of speech. The subsequent two days, his difficulty in swallowing and a nasogastric tube was inserted. You can see that there is a neurological illness which has developed and it is progressing very faster and seems to have ascended from below.

In the ICU, doctors noticed a drop in the saturation to 94 percentage in room air. Patient was tachypneic. On examination, the respiratory system wise all are normal. His single breath count is less.

It is only 10. ABG shows a decrease in PAO2 and also an increase in the PACO2. So, this comes under what is called as Type2 respiratory failure or hypercapnic respiratory failure, which is the classical thing which occurs in restrictive lung disease. So this is a case of restrictive lung disease where the problem is not in the parenchyma, problem is not in the pleura, but then the problem is in the neuromuscular, that is, it could be here in this case it is a problem in the nerve like Guillain-Barre syndrome. Now we are coming

to the end of the session. Now to summarize, respiratory diseases are broadly classified into three types.

Obstructive, where the problem is in the airway. Obstructive where we know the problem can be in the parenchyma, pleura or the neuromuscular or pulmonary vascular where the problem is in the pulmonary vasculature. Now restrictive lung disease, the topic we are discussing, there are, it is classified into three types. One is the problem is parenchyma, pleural and neuromuscular. The commonality among all the restrictive lung disease is that there is a limitation in the ability of the lung to expand during inspiration

That we call it as decreased distance ability or compliance or the elasticity of the lung. Then this is diagnosed by spirometry which shows that the total lung capacity and the functional, forced vital capacity are reduced. Now diffuse parenchyma lung disease of the three restrictive lung disease, diffuse parenchyma lung disease is also called as interstitial lung disease. Interstitial lung disease is classified based on whether it is known cause or unknown cause. Among the known cause we have got environmental exposure to dust like silica and asbestos, drug exposure, secondary to connective tissue disorders and there is another thing called idiopathic where the etiology is not known but commonly associated with smoking. Among the ILD of unknown cause, IPF, idiopathic pulmonary fibrosis is the commonest and it occurs in elderly smoker's male 50 to 60 years of age and with the classical, the ground that is the honeycombing in the sub pleural areas.

Now the cardinal symptom of any restrictive lung disease is dyspnea which is exertional and slowly progressive and dry cough. The classical examination finding to diagnose a diffuse parenchyma lung disease ILD is the presence of Velcro crepitations which is fine end inspiratory crepts and it is heard at the infraaxillary, infra-scapular and interscapular areas. With this we conclude this presentation. Thank you for your patient listening.

Thank you.