Pulmonary Function tests - Interpretation and application in clinical practice

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Week 4
Lecture 2

Interpretation of pulmonary function tests in Restrictive lung disease

Dear friends, welcoming you all for this session on interpretation of pulmonary function test in restrictive lung diseases. What are restrictive lung diseases? Restrictive lung diseases are a heterogeneous set of pulmonary disorders defined by restrictive patterns on spirometry. These disorders are characterized by a reduced distensibility of the lungs compromising lung expansion and in turn reduced lung volumes particularly with reduced total lung capacity. What are its types and causes? Restrictive lung diseases can be caused by pulmonary parenchymal diseases which are labelled as intrinsic causes and extra pulmonary diseases which can be labelled as extensive causes. Intrinsic or pulmonary causes involve the lung parenchyma itself while the extrinsic restrictive lung diseases originate from neuromuscular disorders, obesity and other extra parenchymal disorders like pleural pathologies. In both intrinsic or extrinsic pulmonary conditions lung volumes become reduced due to restrictions in pulmonary mechanics.

Causes for restrictive lung diseases you can use the acronym PAINT, pleural, alveolar, interstitial, neuromuscular and thoracic cage abnormalities. Among these five, alveolar and interstitial constitute the intrinsic causes, rest of the three causes are extrinsic causes. This diagram depicts the differences or different causes in intrinsic and extrinsic pathology. Parenchymal causes which are intrinsic includes interstitial pulmonary fibrosis, occupational lung diseases, collagen vascular diseases, granulomatous diseases, post radiation fibrosis, post pneumonectomy or resection and drug induced pulmonary fibrosis etcetera.

Whereas, extra pulmonary can be classified under pleural chest wall or abdominal causes, pleural causes include pleural effusion, pneumothorax, pleural fibrosis, pleural tumors and pleural thickening. Chest wall based causes include trauma, congenital chest wall abnormalities like kyphosis, scoliosis, ankylosing spondylitis, neuromuscular disease like myasthenia or Gullian Barre, morbid obesity etcetera. And the abdominal causes includes any condition which causes increase in the width of the abdomen and upward displacement of the diaphragm which happens in massive ascites or any condition which causes obstruction within the abdomen and distension of the abdomen. What are the PFT patterns

in restrictive lung diseases? Restrictive abnormalities, restrictive ventilated effects are characterized by a normal or increased FEV1 FVC ratio which is more than 70 percent and a reduction in TLC below 5th percentile or 80 percent of the predicted value. They can be suspected if the FVC is reduced that is less than 80 percent, FEV1 by FVC is normal or increased that is more than 70 percent and the flow volume curve shows a convex shape.

The two types of restrictive abnormalities can be differentiated based on the residual volume and residual volume TLC ratio. However, a reduced forced vital capacity by itself does not prove a restrictive ventilator impairment. Indeed it is associated with a low total lung capacity less than half the time. Conversely in adults a normal forced vital capacity and the FEV1 bar FVC ratio are highly reliable at ruling out restriction as measured by low total lung capacity. If you see the time volume curve this is the normal pattern what we see, but in obstructive pattern and you get a steep decline and in restrictive pattern you can get a convexity instead of the added concavity.

This can be easily looked upon in the flow volume loop. And if you see the time volume curve there is a symmetrical reduction in the FEV1 as well as FVC. So, the ratio is preserved, but if you see the curve it is totally decreased in height as well as the time factor. How to differentiate between obstructive and restrictive pattern? If you see this chart this will help us with the following factors. Forced vital capacity is decreased or normal in obstructive pattern whereas, it is decreased in restrictive.

And FEV1 is always decreased in obstructive pattern and it is decreased or normal in restrictive pattern. The most important factor is FEV1 FVC ratio which is decreased in obstructive pattern which is normal in restrictive pattern. And the total lung capacity is normal or increased in obstructive pattern whereas, in restrictive pattern it is decreased. And as we have already told about the flow volume loop in restrictive lung function, in a restrictive lung function there is a steep slope and a decreased volume and here in obstructive there is a scoop out with a decreased slope with a concavity.

And if you see the time volume curve it parallels the normal tracing with lesser amplitude in restrictive whereas, in obstructive it is prolonged because it takes longer time to get it expelled out. And if you see the volumes and capacities the middle one shows the residual volume, expiratory reserve volume, tidal volume and inspiratory reserve volume. With reference to these all the parameters are reduced in restrictive lung diseases. RV is reduced, expiratory reserve volume is reduced, tidal volume is less and IRV is also reduced. When compared with the normal there is a significant increase in the residual volume, ERV is less than normal, tidal volume is preserved whereas, increased increased inspiratory reserve volume.

So, if you see there is an increase in volume because of the obstruction and retention of the flow because the air is not able to be expelled out. How to differentiate between two types

of restrictive diseases? Parenchymal restrictive disease will show that the total lung capacity is reduced, FEV1 FVC ratio is normal or above normal, residual volume is decreased, there is a low residual volume TLC ratio. Whereas, extraparenchymal restrictive disease total lung capacity is reduced, FEV1 FVC ratio is normal or above normal, residual volume is normal if it is due to muscle weakness or pleural disease, residual volume may be increased if it is due to deformed chest wall. There is a high residual volume TLC ratio, this differentiates between the parenchymal causes for restrictive lung function and extra parenchymal causes for restrictive lung function. And this chart shows the various possibilities with various conditions like pulmonary fibrosis, neuromuscular diseases, severe obesity etcetera.

So, there is a varying degrees of variation in the residual volume and there is a increase in the residual volume, there is a increase in the residual volume in severe obesity, neuromuscular diseases whereas, in pulmonary fibrosis it is reduced when compared with the normal people. What is complex restriction? FEV1 FVC and TLC are typically reduced in roughly the same proportion, this pattern is known as simple restriction. However, some individuals present with a reduction in FVC that is out of proportion to the reduction in total lung capacity indicating a disproportionately elevated residual volume. This pattern is termed complex restriction and is associated with processes that impair lung emptying such as neuromuscular disease, chest wall restriction or occult obstruction with gas trapping. What are the algorithms which can help us to interpret the spirometry? This is one of the recommended algorithm in which the FEV1 FVC ratio is the criteria.

If you see the FEV1 FVC ratio, this is the criteria for the FVC. If it is less than 70 percent of the GOLD criteria or if it is less than the lower limit of normal as per 80s criteria in the adults and in the 5 to 18 years of age if it is less than 80 percent 85 percent predicted, then we have to check the FVC. Then if the FVC is less than the lower limit of normal in the adults as well as in the 5 to 18 years age less than 80 percent of predicted, then it shows mixed pattern. If it is not, then it shows obstructive pattern. So, then you grade the severity according to the bronchodilator reversibility and find out whether the patient has got a reversible obstruction or irreversible obstruction.

When there is a mixed pattern, once again we will be checking with the bronchodilator therapy to find out whether it is a pure obstruction with the air trapping or secondary to chronic obstructive pulmonary disease. On the contrary, there is no reduction. Then you check the FVC whether the FVC is less than lower limit of normal in adults and less than 80 percent, then it is considered as a restrictive pattern and then you assess the grade for severity. When there is a restrictive defect, consider the differential diagnosis whether it is due to parenchymal or extra parenchymal causes. This algorithm gives an alternative approach where the FEV1 FVC ratio is taken as a criteria.

It can be normal or high, otherwise it can be low also. When it is normal or high, see the vital capacity or FVC. If the vital capacity or FVC is normal, it is considered as a normal spirometry. Do a DLCO.

The DLCO is normal. The pulmonary function is considered as normal. On the contrary, the DLCO is low, it can be due to parenchymal or vascular disorder. Otherwise, when the FEV1 FVC ratio is normal or high, when the VC or FVC is low, check for the total lung capacity. If the total lung capacity is low, there is a possibility that could be due to restrictive lung disease. Then check with the DLCO.

If the DLCO is also low, then it could be due to parenchymal causes. If the DLCO is normal, then whether there is any chest wall limitation, we have to cross check. When the vital capacity or FVC is low, at the same time total lung capacity is normal, it could specify non-specific pattern, whether it is due to airway resistance, we have to check. On the contrary, when the FEV1 FVC ratio is low, check the vital capacity or forced vital capacity. If it is low, check the total lung capacity.

If the total lung capacity is normal, that shows it could be obstruction with air trapping. Or if the total lung capacity is high, then it could be due to obstruction with over hyperinflation. Or if the total lung capacity is low, it could be due to mixed pattern. So, here and this type of situation go for bronchodilator reversibility, grade the severity, go for DLCO evaluation. If the DLCO is normal or high, it suggests asthma.

Whereas, if the DLCO is low, it could be due to emphysema, where there is alveolar destruction and it could reflect the emphysema emphysema pattern with volume increase. This is an alternative approach in which the FEV1 bar vital capacity, FEV1 divided by vital capacity is more than or equivalent to vital capacity. If it is yes, then you have to check the vital capacity, whether it is more than or equivalent to the lower limit of normal. If it is more than or equivalent to the lower limit of normal, it is considered as normal. Do DLCO, the DLCO is normal, then it is considered as normal spirometry.

If the DLCO is not normal, it is may be due to pulmonary vascular disorders. On the contrary, if the vital capacity is not more than the lower limit of normal, then check with the total lung capacity, find out whether it is more than the lower limit of normal. If it is more than the lower limit of normal, then there is evidence of obstructive airway disease. On the contrary, if it is lower than the normal limits, then it could be due to restrictive pattern. Then do a DLCO, make sure whether it is due to parenchymal restrictive causes or whether it is extra parenchymal causes.

And in obstruction, you have to find out whether there is a mixed defect or obstruction per se. Do a DLCO and then find out if the DLCO is more than the lower limit of normal, it could be due to asthma and chronic bronchitis. Whereas, if the DLCO is reduced with the obstructive pulmonary function, it will be due to emphysema. How will you assess the

severity of restrictive lung diseases? This is based on the ATS guidelines, which says if the total lung capacity is less than 80 percent, we can grade it as follows. If it is 70 to 80 percent of predicted, it could be mild restrictive disease.

If it is 60 to 70 percent of predicted, it could be moderate restrictive disease. If it is 50 to 60 percent of predicted, it could be moderately severe restrictive disease. And if it is less than 50 percent of predicted, it could be due to severe restrictive disease. How we can use DLCO to assess the restrictive lung function? Full PFT with the DLCO provide the patient's total lung capacity. The restrictive pattern is confirmed as a true restrictive defect, if the total lung capacity is less than 80 percent of the predicted in patients 5 to 18 years of age or less than the lower limit of normal in adults.

So, based on the DLCO, if the DLCO is high, that is more than 140 percent predicted, the DLCO will be labelled as normal if it is 76 to 140 percent. It will be labelled as mild decrease if it is 61 to 75 percent. It will be labelled as moderate decrease if it is 40 to 60 percent. And if it is less than 40 percent, it will be labelled as severe decrease in the DLCO. Based on the DLCO, how to interpret the pulmonary function? There is a high DLCO.

The scenarios can be asthma, left to right intracardiac shunts, polycythemia or pulmonary alveolar hemorrhage. Whereas, there is a normal DLCO with restrictive pattern, it could be due to extrinsic causes like kypho scoliosis, morbid obesity, neuromuscular weakness and pleural effusion. There is a normal DLCO with the obstructive component, then think of alpha 1 antitrypsin deficiency, bronchial asthma, bronchitis, chronic bronchitis. There is a low DLCO and there is a restrictive lung function in the PFT. Think of conditions like asbestosis, berylliosis, wherever there is a parenchymal lung damage.

When there is a low DLCO with obstructive pattern in spirometry, think of conditions like cystic fibrosis, emphysema and silicosis, early silicosis. If the DLCO is low with the normal PFT results, think of vascular causes, chronic pulmonary emboli, congestive heart failure, connective tissue diseases, for dermatomyositis polymyositis, early stages of ILD, pulmonary hypertension etcetera. Then we have to understand the importance of KCO which is that is a corrected DLCO to alveolar volume. DLCO divided by alveolar volume determines whether the currently available alveolar spaces are functioning normally. Low DLCO with the low KCO, it is seen in ILD, but in ILD, KCO may be normal also due to patchy involvement.

Low DLCO with normal or high KCO indicates that remaining lung tissue is functioning normally. Say for example, pneumonectomy the residual lung can over expand to compensate. There is a elevated DLCO when compared with alveolar volume, it is increased blood flow in the remaining lung. Then we can compare with the different patterns and different clinical conditions. Say for example, in pulmonary fibrosis the

DLCO is reduced, KCO is preserved normal, alveolar volume is reduced, mechanism is due to alveolitis.

Whereas in neuromuscular diseases the DLCO is reduced. KCO is high, but volume of the alveoli is reduced it is because of the lack of alveolar expansion due to extrinsic causes. Under pulmonary vascular disease, DLCO is reduced, KCO is also reduced, but alveolar volume is preserved normal. This is due to microvascular involvement. Post pneumonectomy, DLCO is reduced, KCO can be high normal, alveolar volume is reduced because there is loss of lung volume post resection, it is because of the loss of alveolar units.

What is pseudo restrictions? In certain patients with obstruction there may be reduced FVC due to air trapping leading to reduced FEV1 FVC ratio giving the appearance of restrictive lung disease. However, the TLC is increased and thus there is no restriction. The smaller FVC is due to air trapping resulting in an increased residual volume. So, this condition we should be in a position to identify when compared with normal spirometry. To summarize full pulmonary function testing including DLCO is required for the evaluation of restrictive pulmonary function abnormalities.

Background clinical details of the patient and imaging report or films will help us for the proper interpretation of restrictive pulmonary function abnormalities. Spirometry, DLCO etcetera should be performed as per specified protocols for obtaining reliable values and proper interpretation of pulmonary function abnormalities. Thank you very much.