

BREATHLESSNESS & RESPIRATORY DISEASES

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INTRODUCTION

Breathlessness is an easily appreciated symptom even by a lay person. Breathlessness or dyspnoea is a common symptom of respiratory and cardiac diseases although it may occur in other conditions such as renal failure, anaemia, diabetic ketoacidosis, fever, etc.

The term dyspnoea has a Greek origin which translates into (dys) difficult (pnoea) breathing. However, the symptom of breathlessness has a very complex subjective interpretation and description almost to the point of confusion for the clinician.

Breathlessness is a very important and sometimes the only symptom of respiratory disease. Although no definition of dyspnoea is totally satisfactory; an 'unpleasant subjective awareness of the sensation of breathing' is accepted universally.

A Case of Anxious Housewife

Mrs. Sharma, a housewife aged 50 years, non-smoker, non-diabetic and non-hypertensive presented to her family doctor with breathlessness on exertion. Mrs. Sharma's usual exercise was to walk a kilometer to the local vegetable market on alternate evenings. Over the past 3 months, she noticed heaviness and tightness of chest after 3/4 of a kilometer. The routine lab parameters of CBP, RBS, CUE, ECG were normal. The family doctor was not satisfied with Mrs. Sharma's Chest X-ray. It was a poor quality film as the patient was obese and the film was taken during expiratory phase. The family doctor knew Mrs. Sharma's family well and was aware that Mrs. Sharma was unduly anxious about her unmarried grown up children. A course of anxiolytics for 2 weeks did not help the situation. She was referred to the nearby

pulmonologist who noticed that Mrs. Sharma's respiratory rate was 25 per minute and she had clubbing. A repeat X-ray of chest in deep inspiration with a slightly higher KV recorded fine reticular patterns at the lower zones. On auscultation, fine end inspiratory (velcro) crackles were detected at the bases of both the lungs. A spirometry was done which showed a low (60% predicted) FVC (Forced Vital Capacity), a good (90%) (Forced Expiratory Volume₁) FEV₁/FVC ratio. The room air oxygen saturation measured by pulse oxymeter was 88%.

Grades of Breathlessness

After Medical Research Council

- | | |
|---------|---|
| Grade 1 | Troubled by shortness of breath when hurrying on level ground or walking up a slight hill |
| Grade 2 | Short of breath when walking at own pace on level ground |

After Sherwood Jones

- | | |
|----------|--|
| Grade 1a | Able to do housework or job with moderate difficulty |
| Grade 1b | Carrying out job or housework with great difficulty |
| Grade 2a | Confined to chair or bed but able to get up with moderate difficulty |
| Grade 2b | Confined to chair or bed but able to get up with great difficulty |
| Grade 3 | Totally confined to chair or bed |
| Grade 4 | Moribund |

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The New York Heart Association functional and therapeutic classification applied to dyspnoea

Grade 1	No breathlessness
Grade 2	Breathlessness on severe exertion
Grade 3	Breathlessness on mild exertion
Grade 4	Breathlessness at rest

Causes of Breathlessness : Although 'Breathlessness' can be classified by various methods, the best method is to approach anatomically; that is diseases of the airway, diseases of lung parenchyma and diseases of the pleura and chest wall.

Diseases of the airway usually produce cough with or without expectoration. Asthma, Chronic Obstructive Pulmonary Disease (COPD) are the usual causes. breathlessness is episodic and sometimes seasonal in asthma while in COPD, it is progressive and has occasional peaks (3 or 4) per year. There is an associated wheeze or rhonchi on auscultation. X-ray of chest is usually unremarkable and may sometimes show dark (air trapped) lungs with low and flat diaphragms and narrow heart. The lung function test (spirometry) is usually diagnostic. The FEV₁ is reduced and the ratio to FVC also is reduced thereby signifying obstruction. Hypoxia (SpO₂ < 90%) is a very late feature of severe airways disease.

Diseases that affect lung parenchyma such as diffuse infiltrative diseases (sarcoidosis), fibrosing alveolitis, alveolar cell carcinoma) cause progressive breathlessness. Chest X-ray shows reticular and nodular diffuse bilateral opacities, and spirometry typically shows a reduced forced vital capacity. Pneumocystis Carinii Pneumonia in HIV infected patients is an increasingly common parenchymal manifestation in the last 10 years.

Diseases of the pulmonary circulation such as vasculitis or primary pulmonary hypertension cause hypoxia and a severe shunt. Pulmonary thrombo embolism is a classic example where dyspnoea and hypoxia are of a sudden onset while in a chronic state produce polycythemia and cor pulmonale.

Varied types of pleural and chest wall diseases can produce dyspnoea. Pneumothorax, pleural effusion are examples of acute conditions while kyphoscoliosis, poliomyelitis, motor neuron disease are examples of a chronic state of breathlessness.

Diagnosis of Mrs. Sharma

Mrs. Sharma had no "airway symptoms like cough, expectoration, wheezing, etc. and her spirometry showed normal FEV₁/FVC ratio¹. The classic combination of dyspnoea, velcro rales, restrictive pattern of spirometry and the ground glass appearance of lung shadows on chest X-ray point towards a diagnosis of interstitial lung disease (ILD). Idiopathic Pulmonary Fibrosis (IPF) also known as Cryptogenic Fibrosing Alveolitis which by far is the commonest ILD occurring in the age group of 50-70 years.

Mrs. Sharma's pulmonologist later asked for a high resolution CT Scan of Chest (HRCT)^{3,4} which showed ground glass appearance and areas of thickened interlobular septa giving a reticular pattern and a few areas of honey combing. This characteristic appearance correlates well with histopathology so much so that it almost obviates the need for a surgical biopsy⁵. The HRCT features of IPF are reasonably characteristic and in the right clinical context, is diagnostic.

The main causes of honeycomb lung

Localized	Diffuse
Systemic Sclerosis	Cryptogenic fibrosing alveolitis
Sarcoidosis	Rheumatoid lung
Tuberculosis	Langerhans' cell histiocytosis
Asbestosis	Tuberous sclerosis
Berylliosis	Neurofibromatosis

In order to assess the physiological severity of the disease, the specialist ordered two more tests apart from spirometry, viz., 1) Six minute walk test and 2) Measurement of diffusing lung capacity Six minute walk distance test where the room air oxygen saturation is measured along with respiratory rate and distance covered in six minutes of self paced walking. Mrs. Sharma's resting PaO₂ of 60 mm Hg corresponding to SpO₂ of 90% dropped to 50 mm Hg (SpO₂ 80%). This happened because she was in the steep part of the oxygen dissociation curve.⁶⁻⁹ and her respiratory rate went up from 25 to 32.

Single breath diffusing capacity performed with carbonmonoxide is reduced early in IPF.^{6,8} The DLCO depends on the ventilation (V_A) and perfusion (Q) as well as area and thickness of the alveolar membrane.

To compensate for the volume of the lung, the DLCo is expressed as (Kco) transfer co-efficient.

Bronchoscopy and broncho alveolar lavage (BAL) have no great role in Mrs. Sharma's case except perhaps to rule out an upper airway tumor in the trachea or larynx. (Such lesions would normally cause cough, stridor and a defective flow volume loop on spirometry). BAL in evaluation of cryptogenic fibrosing alveolitis has little value excepting for prognostic purposes. High (>4%) neutrophils indicate poor response to steroids while high (>11%) lymphocyte indicate good response to steroids.¹⁰

Mrs Sharma's doctor also ordered for serum levels of rheumatoid factor and anti nuclear antibodies (ANA). In many of the connective tissue diseases, the lung is involved and manifests as fibrosing alveolitis.

Rheumatoid factors are immunoglobulins of the IgG, IgA or IgM class. ANA are detected by indirect immunofluorescence and is a good screening test for systemic lupus erythematosus. Both the tests were negative.

Pratice Points

1. Interstitial lung diseases especially idiopathic pulmonary fibrosis is on the rise and should be considered while evaluating a case of breathlessness.
2. Breathlessness associated with finger clubbing, basal ("Velcro") crepitations and classic ground glass appearance virtually confirms the diagnosis of idiopathic pulmonary fibrosis.
3. Surgical lung biopsy and broncho alveolar lavage contribute very little to the final outcome and treatment of the condition.

Varied Expressions of Breathlessness : Breathlessness is expressed differently by people based on gender, race, culture, language spoken, social background, etc. Different terms like chest tightness, suffocation, not getting enough air, difficulty in breathing, etc. are used.¹¹⁻¹³

Breathlessness in interstitial disease: Patients with restrictive physiological pattern have to breathe at low lung volumes. They experience "unsatisfied breath", inspiratory difficulty" and "rapid and shallow" breathing.^{14,15}

A Case of a successful businessman : Mr. Ram Reddy, 65 years, built a successful cloth business and leading a semi retired life. Mr. Reddy presented to his physician with cough and expectoration of 10 days duration. Mr. Reddy also complained of difficulty in breathing even with routine chores like combing hair and putting on a shirt. Mr. Reddy's wife in fact gave more information that the cough, expectoration and breathlessness had been troubling her husband for the past 4 years. The symptoms are present throughout the year and get worse at least 3 times a year. An avid morning walker, Mr. Reddy cut short his walking time from 45 minutes to 14 minutes and for the past one year, is confined to his house. Mr. Reddy is a smoker and is smoking about 5 cigarettes a day for the past one year.

Earlier, he was smoking 40 cigarettes a day for almost 40 years (40 cigarettes = 2 pack x 40 years = 80 pack years). Not a diabetic, Mr. Reddy has been hypertensive for 20 years and has been using a ACE inhibitor. On examination: RR 20 per minute HR 110 Pulse Oxymeter saturation (SpO₂) was 80%, weight: 80 Kgs (Body Mass Index: 28), mild cyanosis, puffy face, dilated forearm veins and distended neck veins. Mr. Reddy has grossly decreased breath sounds and had bilateral extensive polyphonic rhonchi and a few scattered crepitations. Liver was enlarged (3 fingers) soft and tender and had hepatojugular reflux. There was bilateral pitting edema of feet. Flapping tremors were seen. A provisional diagnosis of Chronic Obstructive Pulmonary Disease (COPD) with acute exacerbation and cor pulmonale was made. The reports of the investigations were as follows. Chest X-ray showed mild cardiomegaly with dilated SVC and dilated right atrium, prominent and dilated pulmonary arteries (features of PAH), dark lungs low flat diaphragms (all features of air trapping), blood showed polycythemia. Spirometry showed the classical features of grossly reduced FEV₁ (40% predicted) with absolute value of 0.60 liters. The FEV₁/FVC percentage was 55% with no reversibility seen following bronchodilator. ECG showed P Pulmonale (P wave in Lead II > 2.5mm). 2D Echo confirmed the ECG and Chest X-ray findings of dilated RA and RV and significant pulmonary arterial hypertension. Analysis of arterial blood gases showed compensated respiratory acidosis (pH 7.35) raised PaCO₂ (55 mm Hg) and significant hypoxia (PaO₂=60 mm Hg).

Breathlessness in COPD is always progressive as the FEV₁ in smokers declines at a rate of 60 ml per year (normal 30 ml per year). Breathlessness becomes significant when FEV₁ is less than one litre. Air trapping due to generalised airways obstruction causes breathlessness. Small airways and lung parenchymal destruction cause \dot{V}/\dot{Q} mismatch and is responsible for hypoxia which in turn is responsible for pulmonary arterial hypertension and right heart involvement ("Cor pulmonale") which is evident on clinical examination. Features of hypoxia hypercarbia can be made out clinically in the form of flapping tremors, dilated forearm veins, headache, drowsiness, etc. Mr. Reddy's COPD can be classified as severe (mild FEV₁ 60-79% predicted, moderate FEV₁ 40-59% predicted, severe FEV₁ <40% predicted).

Learning Points

1. COPD is a very common respiratory disease manifesting as breathlessness.
2. Breathlessness can be measured by several clinical indices.
3. Breathlessness in respiratory disease is mostly due to \dot{V}/\dot{Q} mismatch.

Measurement of Breathlessness : Measurement of dyspnoea (breathlessness) is important for purposes of intervention and also for prognostic reasons. Measurement can be both objective and subjective. Borg Visual Analog Scale (VAS) or a category ratio scale may be used to assess dyspnoea during initial assessment and also after treatment¹⁶ on a scale of 10 cm with markings at every 1 cm. The subject is instructed to provide quantification of his/her breathlessness with "0" as no breathlessness and "10" as intolerable breathlessness.

MRC Scale: The Medical Research Council has introduced a simple method in 1959¹⁷ which is based on perceived breathlessness related to performing a task.

Medical Research Council (MRC) Scale

Five statements about perceived breathlessness:

1. I only get breathless with strenuous exercise.
2. I get out of breath when hurrying on the level or up a slight hill.

3. I walk slower than people of the same age on the level because of breathlessness or have to stop for breath when walking at my own pace on the level.
4. I stop for breath after walking 100 yards or after a few minutes on the level.
5. I am too breathless to leave the house.

BDI/TDI : The Baseline (BDI) and Transition (TDI) Dyspnoea indexes are based on functional impairment and also magnitude of effort.¹⁸

CRDQ: Chronic Respiratory Disease Questionnaire is based on four components to measure quality of life. Dyspnoea is one of the components. The individual patient is asked to recollect five most important activities that have caused dyspnoea over the previous two weeks and grade severity over a scale of 1-7 for each of the five activities. Overall total is divided by 5. A list of 26 activities are prompt to guide the patient.¹⁹

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