Progressive shortness of breath

“Case presentation and interpretation”

SUPERVISOR    DR. RAMI AL-HAYALI
BY    DALAL AHMED & THAKER TAHA
History

• **Name:** شيرو مللو شيرو
• **Age:** 70 years old
• **Residence:** كوكجلي
• **Occupation:** Retired
• **Religion:** Muslim
• **D.O.A:** 9-9-2012
• **D.O.E:** 9-9-2012
Chief complaint:

Shortness of breath for 4 years
History of the present illness:

• A 70 years old man who smoked for 25 years (2 packs/day) and stopped smoking 25 years ago was in a good health state until 4 years ago when he started to complain from **gradual** onset of **cough, dry** most of the times with occasional **whitish sputum** with no blood.
C.C: Shortness of breath for 4 years

H.P.I:

- A 70 Y old man who smoked for 25 years (2 packs/day) and stopped smoking 25 years ago was in a good health state until 4 years ago when he started to complain from gradual onset of cough, dry most of the times with occasional whitish sputum with no blood.

- The cough was present throughout the day and night not responding to any medication, sometimes awakes the patient from sleep.
C.C: Shortness of breath for 4 years

H.P.I:

• The cough was present throughout the day and night not responding to any medication, sometimes awakes the patient from sleep.

• Then about 1 year after that he started to develop shortness of breath which was gradual in onset brought on when the patient walks for the same distance he used to walk for shopping (about 1km), relieved by rest, not associated with dyspnea on lying flat or P.N.D but with persistence of cough.
C.C: Shortness of breath for 4 years

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• Then about 1 year after that he started to develop shortness of breath which was gradual in onset brought on when the patient walks for the same distance he used to walk for shopping (about 1km), relieved by rest, not associated with dyspnea on lying flat or P.N.D but with persistence of cough.

• Then during the 3rd year, the shortness of breath increased in severity and occurred when patient goes to mosque which is **50 meters** away. The cough increased in severity with no noisy breathing, no chest pain, no palpitation, and no leg swelling.
Review of other systems:

• **Locomotor**: low back pain, no joint pain, no swelling, no stiffness, no skin rash.

• **G.I.T**: -ve

• **Renal system**: -ve

• **C.N.S**: -ve
• **Past medical history:**
  The patient denied chronic diseases including asthma, hypertension, diabetes mellitus or CAD
  No history of T.B

• **Past surgical history:**

• **Family history:**
  No similar symptoms in the family, no known respiratory or cardiac disease in the family.

• **Drug history:**
  No chronic drug use prior to the presentation.

• **Social history:**
  Smoker for 25 years 2 packs /day & stopped 25 years ago.
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• **Drug history:**
  No chronic drug use prior to the presentation.

• **Social history:**
  Smoker for 25 years 2 packs/day & stopped 25 years ago.
**Physical examination**

General examination:
Physical examination

General examination:

• An old man who is conscious and alert, apparently kyphotic, mildly breathless and tachypnic.

• He is **centrally cyanosed**, with telangiectasia on the cheeks and corneal arcus.
Physical examination

General examination:

• Hands:
Physical examination

General examination:

• Hands:
  
  **warm**, cyanosed with palmer erythema and **clubbing** of fingers.

  **no asterixis**.
Physical examination

General examination:

• Hands:
  - warm, cyanosed with palmer erythema and clubbing of fingers.
  - no asterixis.
Physical examination

General examination:

• No leg oedema

• Normal JVP, no goiter, no lymphadenopathy, skin rash, or jaundice.
Physical examination

Vital signs:

• Pulse: 64 BPM, frequent ectopic, bounding large volume.
• Respiratory rate: 32 breath/min
• Blood pressure: 110/70 mmHg
• Temperature: 37°C
Physical examination

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- Blood pressure: 110/70 mmHg
- Temperature: 37°C
Physical examination

Systemic examination:
Physical examination

Systemic examination:

Chest:
Physical examination

Systemic examination:

Chest:

• Inspection
  Moderate degree of kyphosis with mild scoliosis.
  Use of accessory muscles.
  Intercostal and subcostal recession.
  Diminished chest movement.
Physical examination

Systemic examination:

Chest:

• Palpation
  
  Central trachea
  Expansion 2cm symmetrically limited
  Tactile focal fremitus decreased
  Apex beat _ visible apex beat,
      palpable at 4th intercostal space at mid clavicular line,
      thrusting in character.
Physical examination

Systemic examination:

Chest:

• Percussion
  Normal resonant percussion note.
Physical examination

Systemic examination:

Chest:

• Auscultation
  Normal vesicular breathing with prolonged expiratory phase.
  Diffuse, numerous, bilateral end inspiratory crackles.
  High pitch expiratory wheeze.
Physical examination

Systemic examination:

CVS:
Physical examination

Systemic examination:

CVS:
No left parasternal heave
Normal double rhythm
No added sounds
No murmurs
Physical examination

Systemic examination:

Abdomen:
Physical examination

Systemic examination:

Abdomen:

Unremarkable
Physical examination

Systemic examination:

Abdomen:
  Unremarkable

CNS:
Physical examination

Systemic examination:

Abdomen:
Unremarkable

CNS:
Normal cranial nerves
Normal motor and sensory examination.
Differential diagnosis:

• ?
• ?
• ?
• ?
Differential diagnosis:

• Interstitial lung disease

• ?

• ?

• ?

• ?
Differential diagnosis:

- Interstitial lung disease
- Chronic obstructive pulmonary disease
- ?
- ?
- ?
Differential diagnosis:

- Interstitial lung disease
- Chronic obstructive pulmonary disease
- Kyphoscoliosis
- ?
Differential diagnosis:

- Interstitial lung disease
- Chronic obstructive pulmonary disease
- Kyphoscoliosis
- Heart failure
## Investigations

- **Pulmonary function test:**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Observed</th>
<th>Pred.</th>
<th>%Pred.</th>
<th>After Bronchodil</th>
<th>Provocation</th>
<th>% Improvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Forced Vital Capacity (FVC)</td>
<td>1.32</td>
<td>2.82</td>
<td>46.8</td>
<td>1.32</td>
<td>2.00</td>
<td>66.0</td>
</tr>
<tr>
<td>Forced expiratory Volume in 1 second (FEV1)</td>
<td>1.32</td>
<td>2.00</td>
<td>66.0</td>
<td>100</td>
<td>70.92</td>
<td>141</td>
</tr>
<tr>
<td>FEV1/FVC X100</td>
<td>100</td>
<td>70.92</td>
<td>141</td>
<td>100</td>
<td>70.92</td>
<td>141</td>
</tr>
<tr>
<td>Peak Expiratory Flow Rate (PEFR)</td>
<td>5.41</td>
<td>6.43</td>
<td>84.1</td>
<td>5.41</td>
<td>6.43</td>
<td>84.1</td>
</tr>
<tr>
<td>Maximal mid-expiratory Flow rate</td>
<td>3.11</td>
<td>2.52</td>
<td>84.1</td>
<td>3.11</td>
<td>2.52</td>
<td>84.1</td>
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<tr>
<td>MVV</td>
<td></td>
<td></td>
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<tr>
<td>Airway Resistance</td>
<td></td>
<td></td>
<td></td>
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<td></td>
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<tr>
<td>SpO2</td>
<td>98%</td>
<td></td>
<td></td>
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<tr>
<td>PaO2</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>84 mmHg</td>
</tr>
</tbody>
</table>
Investigations

• Pulmonary function test:
• Chest X-ray:
Investigations

- Pulmonary function
- Chest X-ray:
- CT scan:
Investigations

- Pulmonary function
- Chest X-ray:
- CT scan:
Investigations

• Pulmonary function
• Chest X-ray:
• CT scan:
Investigations

- **Pulmonary function test:**
- **Chest X-ray:**
- **CT scan:**
- **Echocardiography:**

**LEFT VENTRICULAR MEASUREMENTS:**

<table>
<thead>
<tr>
<th></th>
<th>patient</th>
<th>normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>End Diastolic Diameter:</td>
<td>45mm</td>
<td>mm</td>
</tr>
<tr>
<td>End Systolic Diameter:</td>
<td>32mm</td>
<td>mm</td>
</tr>
<tr>
<td>IVS Thickness:</td>
<td>11mm</td>
<td>mm</td>
</tr>
<tr>
<td>Left atrial diameter</td>
<td>30mm</td>
<td>mm</td>
</tr>
<tr>
<td>Aortic root diameter</td>
<td>24mm</td>
<td>mm</td>
</tr>
<tr>
<td>Ejection fraction</td>
<td>60%</td>
<td>%</td>
</tr>
</tbody>
</table>

**REPORT:**

1. Left ventricle is normal in size and function. The right ventricle is dilated slightly with increased end systolic pressure to 40 mmHg.

2. Mitral and aortic valves are normal

3. Left atrium is normal

4. Other cardiac chambers and valves are normal

**CONCLUSION:**

MILD PULMONARY HYPERTENSION
Diagnosis:

Idiopathic pulmonary fibrosis (IPF)
Review on IPF

Classification of Idiopathic Interstitial Pneumonias:
Review on IPF

Classification of Idiopathic Interstitial Pneumonias:

- Idiopathic Pulmonary fibrosis (most common)
- Nonspecific Interstitial Pneumonia
- Cryptogenic Organizing Pneumonia
- Desquamative Interstitial Pneumonia
- Acute Interstitial Pneumonia
- Lymphoid Interstitial Pneumonia
Review on IPF

Idiopathic pulmonary fibrosis (IPF)
Review on IPF

**Idiopathic pulmonary fibrosis (IPF)**

Is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, primarily occurring in older adults, limited to the lungs, and associated with the histopathologic and/or radiologic pattern of usual interstitial pneumonia (UIP).
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

Idiopathic pulmonary fibrosis portends a poor prognosis, and, to date, no proven effective therapies are available for the treatment of idiopathic pulmonary fibrosis beyond lung transplantation.
**Review on IPF**

**Idiopathic pulmonary fibrosis (IPF)**

Most patients with idiopathic pulmonary fibrosis present with a gradual onset, often greater than **6 months**, of **dyspnoea** and/or a **non-productive cough**. The symptoms often precede the diagnosis by a median of 1-2 years.
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

It is now believed that epithelial injury and activation in fibroblast foci are crucial early events that trigger a cascade of changes leading to reorganization of pulmonary tissue compartments.
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

The diagnosis of IPF is **definite** in the **presence** of surgical biopsy specimen which show:
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

The diagnosis of IPF is likely in the absence of surgical biopsy specimen with all of the following major criteria and three of minor criteria:
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

• Major criteria
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

- **Major criteria**
  1. Exclusion of other known causes of ILD.
  2. Abnormal pulmonary physiology with evidence of a restrictive process and impaired gas exchange.
  3. Bibasilar reticular abnormalities with minimal ground glass opacities on HRCT.
  4. Transbronchial lung biopsy or BAL inconsistent with alternative diagnosis.
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

• Minor criteria
Review on IPF

Idiopathic pulmonary fibrosis (IPF)

• Minor criteria
  1. Age >50 years.
  2. Insidious onset of otherwise unexplained dyspnoea on exertion.
  3. Duration of illness >3 months.
Review on IPF

Medical Care
Review on IPF

Medical Care

• The goal of any disease management strategy should include assessment and treatment of comorbid medical conditions.

Common comorbid medical conditions found in patients with idiopathic pulmonary fibrosis (IPF) include chronic obstructive pulmonary disease, obstructive sleep apnea, and gastroesophageal reflux disease. Therefore, if any of these comorbid illnesses are present, they should be managed according to current practice guidelines.

Does IPF cause wheeze???
Review on IPF

Medical Care

• **Stop smoking**.

• Patients with hypoxemia (PaO2 < 55 mmHg or oxygen saturation as measured using pulse oximetry [SpO2] < 88%) at rest or with exercise should be prescribed oxygen therapy to maintain a saturation of at least 90% at rest, with sleep, and with exertion.

• **Vaccination** against influenza and pneumococcal infection should be encouraged in all patients with idiopathic pulmonary fibrosis.
Review on IPF

Medical Care

Medication
Review on IPF

Medical Care

Medication

Corticosteroid (Systemic)

- Corticosteroids have not been evaluated in a randomized, placebo-controlled trial to determine their benefit in treating patients with idiopathic pulmonary fibrosis. Retrospective uncontrolled studies have reported no survival benefits. Latent tuberculosis should be excluded before patients are started on corticosteroid therapy.
Review on IPF

Medical Care

Medication

Corticosteroid (Systemic)

prednisolone

• Prevent or suppress inflammation and immune responses when administered at pharmacological doses include inhibition of leukocyte infiltration at the site of inflammation, interference in the function of mediators of the inflammatory response, and suppression of humoral immune responses.
Review on IPF

• Initial response should occur within 3 months of initiating corticosteroid therapy. Improvement in objective parameters such as HRCT imaging, pulmonary function tests, 6MWT, and/or dyspnea scores should be monitored when deciding if additional therapy with prednisolone is warranted.

Which investigation is important to be done for patient on long term corticosteroid??
Review on IPF

Medical Care

Medication

Corticosteroid

DEXA:

<table>
<thead>
<tr>
<th>Region</th>
<th>BMD (g/cm²)</th>
<th>Young-Adult (%)</th>
<th>T-score</th>
<th>Age-Matched (%</th>
<th>Z-score</th>
<th>BMC (g)</th>
<th>Area (cm²)</th>
<th>Width (cm)</th>
<th>Height (cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>L1</td>
<td>0.907</td>
<td>85</td>
<td>-1.3</td>
<td>95</td>
<td>-0.4</td>
<td>9.81</td>
<td>10.81</td>
<td>4.0</td>
<td>2.62</td>
</tr>
<tr>
<td>L2</td>
<td>0.930</td>
<td>73</td>
<td>-2.6</td>
<td>81</td>
<td>-1.6</td>
<td>7.72</td>
<td>9.30</td>
<td>3.9</td>
<td>2.48</td>
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<tr>
<td>L3</td>
<td>0.737</td>
<td>64</td>
<td>-3.4</td>
<td>71</td>
<td>-2.6</td>
<td>8.68</td>
<td>11.79</td>
<td>4.1</td>
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<tr>
<td>L4</td>
<td>0.649</td>
<td>57</td>
<td>-4.1</td>
<td>62</td>
<td>-3.3</td>
<td>8.33</td>
<td>12.84</td>
<td>4.6</td>
<td>5.00</td>
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<tr>
<td>L1-L2</td>
<td>0.872</td>
<td>80</td>
<td>-1.9</td>
<td>89</td>
<td>-0.9</td>
<td>17.52</td>
<td>20.10</td>
<td>4.0</td>
<td>7.99</td>
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<tr>
<td>L1-L3</td>
<td>0.822</td>
<td>74</td>
<td>-2.5</td>
<td>82</td>
<td>-1.5</td>
<td>26.21</td>
<td>31.89</td>
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<td>7.99</td>
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<tr>
<td>L1-L4</td>
<td>0.772</td>
<td>68</td>
<td>-3.0</td>
<td>76</td>
<td>-2.0</td>
<td>34.54</td>
<td>44.73</td>
<td>4.1</td>
<td>10.76</td>
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<tr>
<td>L2-L3</td>
<td>0.778</td>
<td>68</td>
<td>-3.1</td>
<td>75</td>
<td>-2.1</td>
<td>16.40</td>
<td>21.08</td>
<td>4.0</td>
<td>5.28</td>
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<tr>
<td>L2-L4</td>
<td>0.729</td>
<td>64</td>
<td>-3.5</td>
<td>70</td>
<td>-2.7</td>
<td>24.74</td>
<td>33.93</td>
<td>4.2</td>
<td>8.03</td>
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<tr>
<td>L3-L4</td>
<td>0.691</td>
<td>60</td>
<td>-3.8</td>
<td>65</td>
<td>-3.0</td>
<td>17.02</td>
<td>24.63</td>
<td>4.3</td>
<td>5.65</td>
</tr>
</tbody>
</table>

T-score for Vertebral Height (L2-L4):
- Compared to young adult (T-score): -6.03
- Adjusted for stature (T-score): -3.79
Review on IPF

Medical Care

Medication

Immunosuppressant Agent
Review on IPF

Medical Care

Medication

Immunosuppressant Agent

**Azathioprine (Imuran)**

- Effects may decrease proliferation of immune cells and result in lower autoimmune activity.
Review on IPF

Medical Care

Medication

Immunosuppressant Agent

Cyclophosphamide

• Prodrug that requires hepatic activation in order to be cytotoxic. Also has immunosuppressant effects. Causes lymphopenia (both B and T cells) and selective suppression of B-lymphocyte activity.
Review on IPF

Medical Care
Medication
Antioxidants
Review on IPF

Medical Care

Medication

Antioxidants

N-acetylcysteine

• An oxidant-antioxidant imbalance may contribute to the pathogenesis of idiopathic pulmonary fibrosis.

• N-acetylcysteine is an antioxidants that may restore balance and would slow functional deterioration in patients with idiopathic pulmonary fibrosis.
Review on IPF

Medical Care

Medication

Antioxidants

N-acetylcysteine

- In spite of NAC, added to prednisone and azathioprine, significantly slowed the rate of deterioration of vital capacity and DLCO at 12 months. However, this did not translate into a survival benefit. Additionally, a significantly lower rate of myelotoxic effects was noted in the group taking NAC so evidence-based guidelines recommend that the majority of patients with IPF should not be treated with N-acetylcysteine monotherapy, however, this therapy may be a reasonable choice in a minority of patients.
Review on IPF

Medical Care

Medication

Antifibrotic agents
Review on IPF

Medical Care

Medication

Antifibrotic agents

Pirfenidone

- A novel compound with combined anti-inflammatory, antioxidant, and antifibrotic effects, had potential therapeutic benefits for idiopathic pulmonary fibrosis.

- Pirfenidone reduced the proportion of patients with a 10% or more decline in FVC by 30% compared with placebo.
Review on IPF

Medical Care

Medication

Antifibrotic agents

Pirfenidone

- Pirfenidone has not yet been approved by the FDA for treatment of IPF; however, the FDA has requested that a new clinical trial be completed. With the new data available, pirfenidone has a favorable benefit-risk profile and represents a potential treatment option for patients with IPF.
Review on IPF

Medical Care

Medication

Antifibrotic agents

Pirfenidone

• Adverse effects

1. Gastrointestinal
   – Nausea, vomiting, dyspepsia, gastritis and gastroesophageal reflux disease (GERD).
   – To reduce the severity of these reactions, pirfenidone is to be taken after meals.
Review on IPF

Medical Care

Medication

Antifibrotic agents

Pirfenidone

• Adverse effects

2. Skin

– Photosensitivity reactions, rash, pruritus and dry skin.
– Patients are usually advised to avoid direct exposure to sunlight, and to use protective clothing and sunscreen agents.
Review on IPF

Medical Care

Medication

Antifibrotic agents

Pirfenidone

• Adverse effects

3. Hepatic dysfunction
   – Increase hepatic enzyme levels, especially those of (AST), (ALT) and (GGT).
   – The drug is contraindicated in patients who have severe hepatic impairment.
Review on IPF

Medical Care

Medication

Antifibrotic agents

Pirfenidone

• Adverse effects

4. Dizziness and fatigue

5. Weight loss
Review on IPF

Medical Care

Medication

Unproven drugs

- Biological response modulators as Interferon-γ1b and Etanercept
- Endothelin receptor antagonists as Bosentan
- Phosphodiesterase inhibitors as Sildenafil
- Tyrosine kinase inhibitors as Imatinib mesylate
- Colchicine
- Anticoagulants as warfarin
Review on IPF

Surgical Care
Review on IPF

Surgical Care

Lung transplantation

• for idiopathic pulmonary fibrosis has been shown to confer a survival benefit over medical therapy.

• Any patient diagnosed with idiopathic pulmonary fibrosis or probable idiopathic pulmonary fibrosis should be referred for lung transplantation evaluation, regardless of the vital capacity.

• Idiopathic pulmonary fibrosis has now replaced chronic obstructive pulmonary disease as the most common indication for lung transplantation in the United States.
Review on IPF

Surgical Care

Lung transplantation

Guidelines for listing a patient with idiopathic pulmonary fibrosis include:

1. Diffusion capacity of carbon monoxide (DL\textsubscript{CO}) less than 39% predicted
2. 10% or greater decrement in forced vital capacity during 6 months of follow-up
3. Decrease in pulse oximetry below 88% during a 6-minute walk test (6MWT)
4. Honeycombing on high-resolution computed tomography (HRCT) imaging (fibrosis score >2).

The reported 5-year survival rates after lung transplantation in idiopathic pulmonary fibrosis are estimated at 50-56%.
Review on IPF

Complications
Review on IPF

Complications

The following are complications that can be seen in patients with idiopathic pulmonary fibrosis:

1. Pulmonary hypertension
2. Acute exacerbation of pulmonary fibrosis
3. Respiratory infection
4. Thromboembolic disease
5. Adverse medication effects
6. Lung cancer
Review on IPF

Mortality/Morbidity
Review on IPF

Mortality/Morbidity

• Idiopathic pulmonary fibrosis portends a poor prognosis, a worse prognosis can be expected based on various clinical parameters, physiologic factors, radiographic findings, histopathologic findings, laboratory findings, and bronchoalveolar lavage findings.

• Estimated mean survival of 2-5 years from the time of diagnosis.

• Death rates in patients with idiopathic pulmonary fibrosis increase with increasing age, are consistently higher in men than women, and experience seasonal variation, with the highest death rates occurring in the winter, even when infectious causes are excluded.
Review on IPF

• **Mortality/Morbidity**

• The most common causes of death in patients with idiopathic pulmonary fibrosis include **acute exacerbations** of idiopathic pulmonary fibrosis, **congestive heart failure**, **lung cancer**, **infectious** causes, and **venous thromboembolic** disease.

• 4 predictors (sex, age, % predicted FVC, and % predicted DL\textsubscript{CO}) could be used in a scoring system to estimate 1-year mortality.
**Review on IPF**

- **Mortality/Morbidity**

Table 1. Scoring for mortality risk in IPF

<table>
<thead>
<tr>
<th>Predictor</th>
<th>Points</th>
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<tbody>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>0</td>
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<tr>
<td>Male</td>
<td>1</td>
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<tr>
<td>Age (years)</td>
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<td>≥60</td>
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<td>61-65</td>
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<td>FVC (% predicted)</td>
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<td>&gt;75</td>
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<td>50-75</td>
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<tr>
<td>&lt; 50</td>
<td>2</td>
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<td>DLCO (% predicted)</td>
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<td>&gt;55</td>
<td>0</td>
</tr>
<tr>
<td>36-55</td>
<td>1</td>
</tr>
<tr>
<td>≤35</td>
<td>2</td>
</tr>
<tr>
<td>Cannot perform</td>
<td>3</td>
</tr>
</tbody>
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Thank you for joining us....

We hope that benefit achieved...