Interstitial Lung Disease (ILD)

Tracy Ward
Highly Specialist Respiratory Nurse – Rotherham NHS Foundation Trust

The views expressed in this presentation are those of the speaker and are not necessarily those of the meeting sponsors. This presentation may contain off-licence information. Please refer to the product SmPCs for the approved indication for use.
Disclosures:

• Teva UK Limited
Managing Idiopathic Pulmonary Fibrosis (IPF)

A ‘can do’ approach
What are Interstitial Lung Diseases (ILDs)?

- A group of >200 respiratory diseases
- Lead to lung fibrosis
- Serious conditions
What are ILDs?

• Some causes are known:
  • Pneumoconiosis

• Some are not:
  • Idiopathic Pulmonary Fibrosis

• Result from damage to the lung by an assault of some kind resulting in:
  • Inflammation
  • Lung fibrosis
Idiopathic Pulmonary Fibrosis (IPF)

- Poor prognosis
- Average survival from diagnosis is 3 years
- 32,500 people in the UK currently diagnosed with IPF
- 5,300 will die within the next 12 months
- If IPF were a cancer it would be the 8th most common
- More people die from IPF each year than from leukaemia
Treatment options: only two!
Anti-fibrotic treatment
pirfenidone and nintedanib

• In some people, proven to slow down the rate of disease progression
• Both drugs now available on the NHS
• Have to meet a prescribing criteria
• Prescribed by regional centres only
• Treatment for IPF ONLY
• Not easily tolerated due to side effects
  – pirfenidone: Gastric disturbance (diarrhoea, nausea, vomiting)
  Photosensitivity
  – nintedanib: Gastric disturbance (diarrhoea, nausea, vomiting)
  Bleeding
Criteria for pirfenidone

• You should be able to have pirfenidone if your Forced Vital Capacity (FVC; a test of your lung function) is between 50% and 80% of the value expected for you

• Pirfenidone should be stopped if your disease gets worse, that is, if your FVC falls by 10% or more in 12 months

• 3 tablets TDS
Criteria for nintedanib

• You should be able to have nintedanib if your Forced Vital Capacity (FVC; a test of your lung function) is between 50% and 80% of the value expected for you.

• Nintedanib should be stopped if your disease gets worse, that is, if your FVC falls by 10% or more in 12 months.

• 1 tablet BD
Transplant

• Most ILD patients referred for transplant have IPF

• Other forms of ILD should be considered for transplant

• Survival rate is 62% at 5 years and 48% at 10 years

Newcastle Hospital Transplant Registry 2004
Transplant

• Strict selection criteria
  – Absolute contraindications
  – Relative contraindications

• Full list can be found at:

• Rigorous assessment by MDT
Disease specific transplant criteria

IPF

• Histologic or radiographic evidence of IPF and any of the following:
  – A 10% or greater decrement in Forced Vital Capacity (FVC) during 6 months of follow-up
  – DLCO of less than 40% with clinical deterioration and/or a greater than 15% decline in DLCO over 6 months of follow-up
  – A rapid decrease in pulse oximetry below 88% during a 6 minute walk test
  – Short rapid decline in symptoms pre-diagnosis

DLCO: Diffusing capacity of the lungs for carbon monoxide
Symptom management
The main symptoms which require management in IPF

- Breathlessness
- Hypoxia
- Cough
- Anxiety
Breathlessness
Pulmonary rehabilitation

• Pulmonary rehabilitation is an effective treatment for breathlessness in chronic respiratory disease

• Assess people with IPF for pulmonary rehabilitation at the time of diagnosis. Assessment may include a 6 minute walk test (distance walked and oxygen saturation measured by pulse oximetry) and a quality of life assessment
Pulmonary rehabilitation

• Pulmonary rehabilitation is part of the NICE management pathway for IPF
• Programme of exercise and education over a period of weeks (usually 12 sessions over 6 weeks)
• Helps people to manage their breathlessness and improve their quality of life
• Should be offered to all patients with ILD who are breathless
Pulmonary rehabilitation breaks the cycle of breathlessness.

1. You feel breathless.
2. You avoid activities that make you breathless.
3. You do less.
4. Your muscles become weaker and less efficient.
5. You get more breathless.

Diagram:
- Arrows from 'You feel breathless' to 'You avoid activities that make you breathless' to 'You do less' to 'Your muscles become weaker and less efficient' to 'You get more breathless.'
Pulmonary rehabilitation

• Treat any other respiratory conditions, ILD may not respond to inhalers but COPD does!
• Use of fans – some patients find that this helps
• Relaxation and breathing techniques
Medication

• Liquid oral morphine* can be very effective for palliating breathlessness. Does not resolve breathlessness but rather ‘takes the edge off’ the feeling of suffocation and makes it more manageable

• 5 - 20mg
• Initially as needed
• Can be used every hour if required

*Liquid oral morphine is indicated for the relief of severe pain in adults, adolescents (aged 13-18 years) and children (aged 1-12 years).
Managing hypoxia with oxygen therapy
What is a normal arterial blood gas?

- The normal pH range is: 7.35 - 7.45
- The normal PaO$_2$ range is: 10.5 - 13.5 kPa (70-100 mmHg)
- The normal PaCO$_2$ range is: 4.7 - 6.0 kPa (35-45 mmHg)
Oxygen therapy

• The requirement for oxygen therapy should be assessed using the British Thoracic Society (BTS) oxygen guidelines

Ambulatory (AOT) or Long Term Oxygen Therapy (LTOT)?

Ambulatory Oxygen (AOT)

• People with ILD will usually require ambulatory oxygen long before they require LTOT

• The requirement for ambulatory oxygen should be assessed at each visit using the ambulatory oxygen guidelines, 6 minute walk test
Long Term Oxygen Therapy (LTOT)

• Assessment should be carried out as per the BTS oxygen guidelines on all patients with resting SaO2 <92%

• Clinical hypoxia is defined as:
  • PaO2 < 7.3 kPa
  • Or
  • PaO2 < 8 kPa with evidence of Pulmonary Hypertension/Heart Failure
Note

• When prescribing oxygen therapy in ILD patients, consideration should be given to any concurrent diagnosis of COPD

• The possibility of Type 2 Respiratory Failure may limit how much oxygen it is safe to give
• In pure ILD:
  Aim $\text{PaO}_2 > 10.5$ with normal PH

• Concurrent COPD
  Aim $\text{PaO}_2 > 8$ with normal PH
Cough
Cough

• One of the most common symptoms affecting people with ILD

• Initially annoying, can become intractable

• Often dry but can be productive if associated with traction bronchiectasis
Cough

Treat causes:
  - GORD
  - Mucus

Medication
  - Mucolytics: carbocisteine
  - Antitussives may help
  - Codeine phosphate tablets 15-30mg QDS*
  - Liquid oral morphine 2.5mg-20mg QDS/PRN*
  - Thalidomide (small studies show this appears to be effective)*

*Not specifically licensed to treat cough
Anxiety

I can't keep calm because I have anxiety.
• Talk to the patient about what is at the route of their anxiety, don’t assume that you know

• Relaxation techniques

• Referral to psychological services may be required:
  – Counselling
  – Cognitive Behavioural Therapy (CBT)
Medication:

• Lorazepam 500mcg S/L PRN (up to 4mg daily)

• Liquid oral morphine 2.5mg-20mg QDS + PRN up to hourly

Liquid oral morphine is indicated for the relief of severe pain in adults, adolescents (aged 13-18 years) and children (aged 1-12 years).
So although we still can’t offer a cure; there is plenty we ‘**CAN DO**’ for these patients.
Any questions?