

Year 6 Integrated Medicine Course:  
Respiratory Lecture 3:  
***Less common lung diseases and  
preparation for PACES***

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# Year 6 Integrated Medicine Course

## Respiratory Lectures

- **Lecture 1:**  
Diagnosis and approaches to the breathless patient
- **Lecture 2:**  
Management of Common Airway Diseases
- **Lecture 3:**  
**Less Common Lung Diseases and preparation for PACES**

Remember : I have not covered respiratory emergencies in these 3 lectures

## Modules produced:

1. A rational approach to the diagnosis of respiratory disease
2. Respiratory Emergencies (including pneumothorax, foreign bodies, Acute asthma, pulmonary embolism, pneumonia and exacerbations of COPD)
3. Asthma
4. Chronic Obstructive Pulmonary Disease
5. Lung Cancer
6. Sarcoidosis
7. Obstructive Sleep Apnoea Syndromes (In preparation)

*PLUS VPAs on TB and severe asthma*

<b>Airways diseases</b>	<b>Small lung disorders</b> (also known as “restrictive disorders”)
<b><i>Localised obstruction</i></b>	<b>Due to disease <i>within</i> the lungs</b>
<ul style="list-style-type: none"> <li>Sleep apnoea</li> <li>Laryngeal carcinoma</li> <li>Thyroid enlargement</li> <li>Vocal cord dysfunction</li> <li>Relapsing Polychondritis</li> <li>Tumours</li> <li>Post tracheostomy stenosis</li> <li>Foreign bodies</li> <li>Bronchopulmonary dysplasia</li> </ul>	<ul style="list-style-type: none"> <li>Sarcoidosis</li> <li>Asbestosis</li> <li>Extrinsic Allergic Alveolitis</li> <li>Fibrosing Alveolitis</li> <li>Eosinophilic pneumonia</li> </ul>
<b><i>Generalised obstruction</i></b>	<b>Due to disease <i>outside</i> the lung</b>
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<b>Infections</b>	<b>Pulmonary vascular disorders</b>
<ul style="list-style-type: none"> <li>Tuberculosis</li> <li>Infective bronchitis</li> <li>Pneumonia</li> <li>Empyema</li> </ul>	<ul style="list-style-type: none"> <li>Pulmonary emboli</li> <li>Pulmonary hypertension</li> </ul>

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In addition to doing the Respiratory Emergencies E module, you are strongly advised to do The VPA on Tuberculosis, and The E module on Lung Cancer

Infections	Pulmonary vascular disorders
<ul style="list-style-type: none"> <li><b>Tuberculosis</b></li> <li>Acute Bacterial Bronchitis</li> <li>Pneumonia</li> <li>Empyema</li> </ul>	<ul style="list-style-type: none"> <li>Pulmonary emboli</li> <li>Pulmonary hypertension</li> </ul>



## Airways diseases

## Small lung disorders (also known as “restrictive disorders”)

### **Localised obstruction**

### **Due to disease *within* the lungs**

Sleep apnoea  
Enlarged cervical lymph nodes  
Thyroid enlargement  
Vocal cord dysfunction  
Relapsing Polychondritis  
Tumours  
Post tracheostomy stenosis  
Foreign bodies  
Bronchopulmonary dysplasia

Sarcoidosis  
Asbestosis  
Extrinsic Allergic Alveolitis  
Fibrosing Alveolitis  
Eosinophilic pneumonia

### **Generalised obstruction**

### **Due to disease *outside* the lung**

Asthma  
COPD  
Bronchiectasis  
Cystic Fibrosis  
Obliterative Bronchiolitis

Pleural effusions  
Pneumothorax  
Scoliosis  
Respiratory muscle weakness  
Obesity

## Infections

## Pulmonary vascular disorders

Tuberculosis  
Infective bronchitis  
Pneumonia  
Empyema

Pulmonary emboli  
Pulmonary hypertension

## Airways diseases

## Small lung disorders

(also known as “restrictive disorders”)

### **Localised obstruction**

### **Due to disease *within* the lungs**

Sleep apnoea  
Enlarged cervical lymph nodes  
Thyroid enlargement  
Vocal cord dysfunction  
Relapsing Polychondritis  
Tumours  
Post tracheostomy stenosis  
Foreign bodies  
Bronchopulmonary dysplasia

Sarcoidosis  
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### **Generalised obstruction**

### **Due to disease *outside* the lung**

Asthma  
C.O.P.D.  
Bronchiectasis  
Cystic Fibrosis  
Obliterative Bronchiolitis

Pleural effusions  
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Scoliosis  
Respiratory muscle weakness  
Obesity

## Infections

## Pulmonary vascular disorders

Tuberculosis  
Infective bronchitis  
Pneumonia  
Empyema

Pulmonary emboli  
Pulmonary hypertension

## Pickwick papers by



“The object that presented itself to the eyes of the astonished clerk, was a boy--a wonderfully fat boy--habited as a serving lad, standing upright on the mat, with his eyes closed as if in sleep. He had never seen such a fat boy, in or out of a travelling caravan; and this, coupled with the calmness and repose of his appearance,.....” Ch 54

## Pickwick papers by



“it was evening. Isabella and Emily had strolled out with Mr. Trundle; the deaf old lady had fallen asleep in her chair; the snoring of the fat boy, penetrated in a low and monotonous sound from the distant kitchen; the buxom servants were lounging at the side door, enjoying the pleasantness of the hour,...”

Ch 8

## Pickwick papers by



'Come along, Sir. Pray, come up,' said the stout gentleman.

'Joe!--damn that boy, he's gone to sleep again.--Joe, let down the steps.' The fat boy rolled slowly off the box, let down the steps, and held the carriage door invitingly open. Mr. Snodgrass and Mr. Winkle came up at the moment.

Ch 4

# Clinical Features of OSAS

- **Snoring**
- **Excessive daytime sleepiness**
- **Impaired concentration**
- Unrefreshing sleep
- Nocturnal choking/wakening
- **Witnessed apnoeas**
- Restless sleep
- Difficulty with concentration
- Impotence
- Nocturia
- Irritability/personality change

# Epidemiology of OSAS

40% of the population snore but what proportion might also have obstructive sleep apnoea syndrome (OSAS)?

# Epidemiology of OSAS

- Up to 5% of adults (M>F) in western countries are likely to have undiagnosed OSA syndrome.

*T. Young et al Am J Respir Crit care Med 2002 165:1217-1239*



# Epidemiology of OSAS: Is there an effect of ethnicity?

- One study of over 65 year old adults showed the OR of significant OSAS was **2.5 times greater for African Americans** relative to Caucasians (controlling for BMI and other confounders)

*(Ancoli-Israel et al Am J Respir Crit Care Med 1995)*

***And there may be other racial and familial predispositions linked to facial morphology or function***

# What predisposes to OSAS?

- Obesity
- Micrognathia
- Macroglossia
- Tonsillar and adenoid hypertrophy
- Mucopolysaccharidoses
- Nasal problems; septal deviation, allergic rhinitis
- Other causes of reduction in calibre of oro-pharyngo-laryngeal airway
- Hypothyroidism, acromegaly
- Downs syndrome
- Polycystic ovarian syndrome
- Sedative drugs
- Alcohol

# Clinical Features of OSAS

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How do we determine what is excessive daytime sleepiness?



The Hammersmith Hospitals NHS Trust  
Sleep Service



**EPWORTH SCALE**

**Name:** \_\_\_\_\_ **Date:** \_\_\_\_\_

**Hospital Number:** \_\_\_\_\_ **Date of Birth:** \_\_\_\_\_

In contrast to just feeling tired, how likely are you to doze off or fall asleep in the following situations.? Even if you have not done some of these things recently, try to work out how they would affect you. Use the following scale to choose the most appropriate number for each situation.

- 0 = no chance of dozing
- 1 = slight chance
- 2 = moderate chance
- 3 = definitely would doze

Situation	Chance of Dozing
Sitting and reading	
Watching T.V.	
Sitting inactive in a public place (e.g. Theatre or a meeting)	
As a passenger in a car for an hour without a break	
Lying down to rest in the afternoon when circumstance permit	
Sitting and talking to someone	
Sitting quietly after lunch without alcohol	
In a car, while stopped for a few minutes in the traffic	

# Differential diagnosis of excessive daytime sleepiness

- Sleep deprivation (Quantity of sleep)
- Sleep fragmentation (Quality of sleep)
- Shift work
- Depression
- Narcolepsy
- Hypothyroidism
- Restless leg syndrome/periodic limb movement disorder
- Drugs including alcohol, sedatives, beta blockers
- Neurological conditions

# Having OSAS leads to

- Impaired Quality of Life
- Disruption of relationships, and
- Day time sleepiness

but there are also other adverse associations.

# Consequences of Obstructive Sleep Apnoea

- Hypoxia
- Hypercapnoea
- Pulmonary Hypertension
- Systemic Hypertension
- Right Heart Failure
- Polycythaemia
- Cardiac Rhythm disturbance
- Sudden death

# Risks associated with Sleep related breathing disorder

- Hypertension
- Cerebro-vascular disease
- Coronary Artery disease
- Road Traffic Incidents
- Occupational Accidents



# Do we diagnose OSAS promptly?

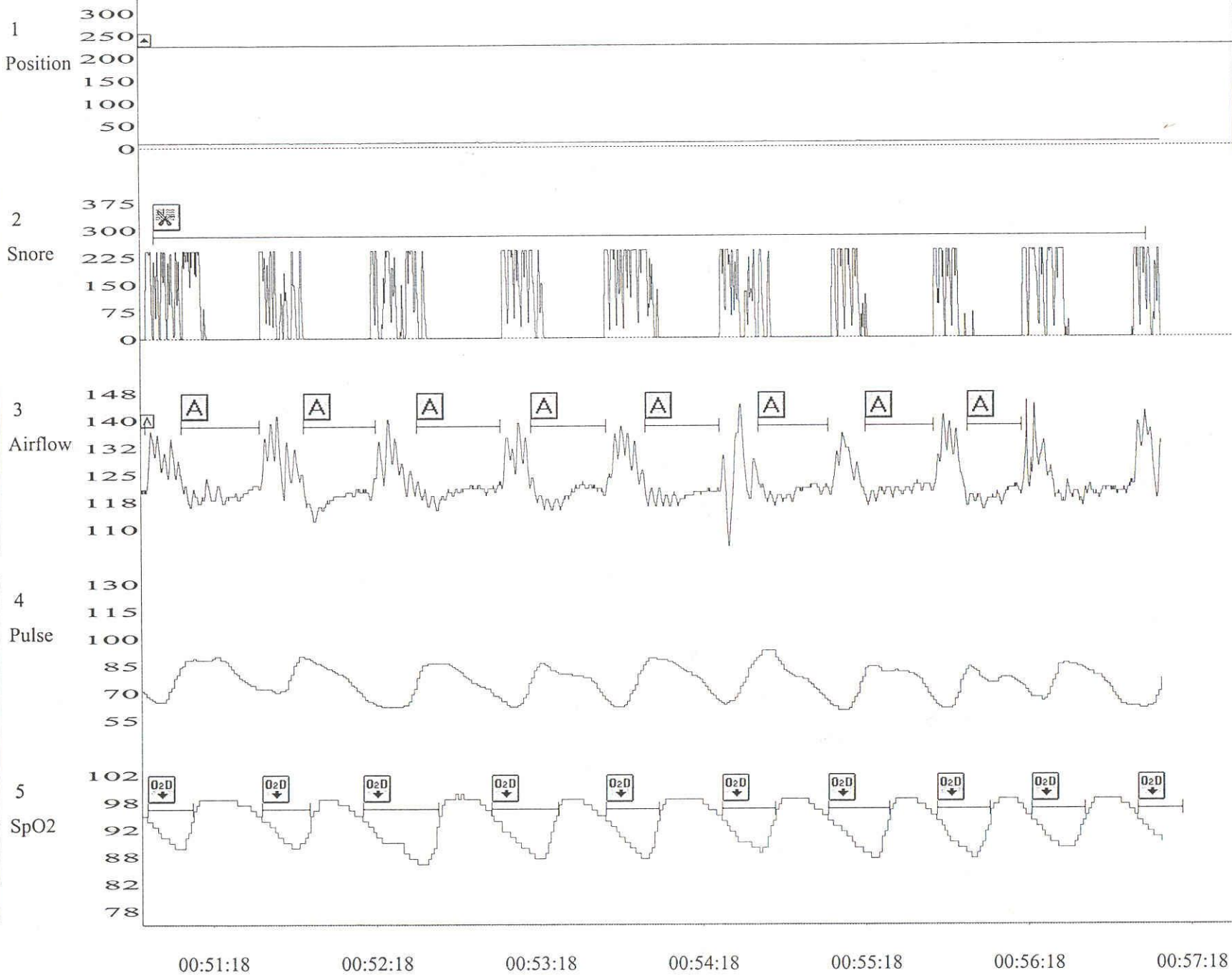
## Duration of probable morbidity prior to diagnosis of OSA

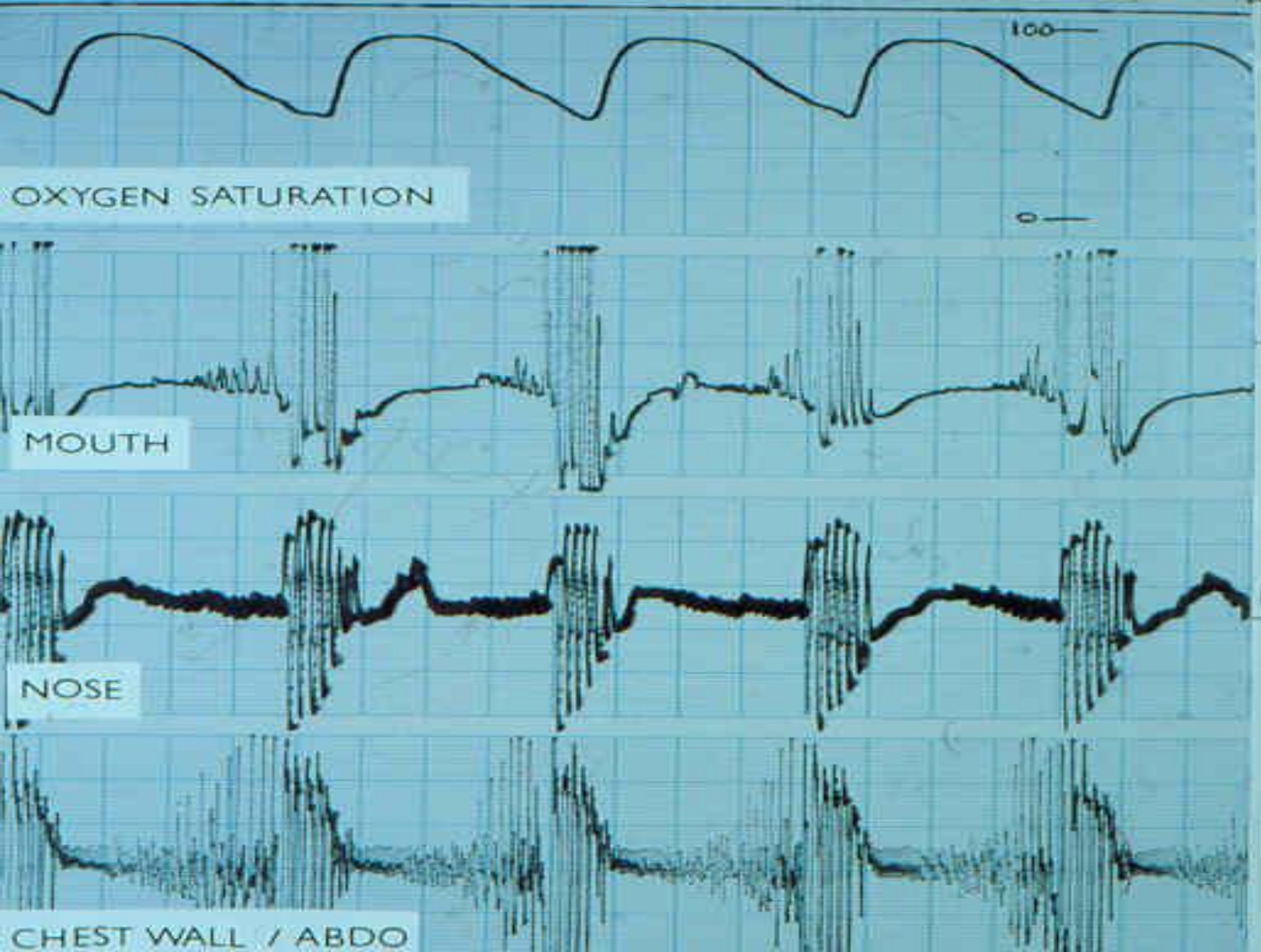
- 117 patients on CPAP treatment for OSAS
- 107/117 (92%) reported complaint of loud **snoring** prior to diagnosis for a **median of 12 years** prior to diagnosis (range 2-47 years)
- 78% reported **witnessed apnoea** for a **median 8 years** prior to diagnosis (range 1- 49 years)
- 83% reported **sleepiness** in the day time for a **median 7 years** prior to diagnosis (range 0.5 – 62 years)

*Ghiassi and Partridge Thorax 2004*

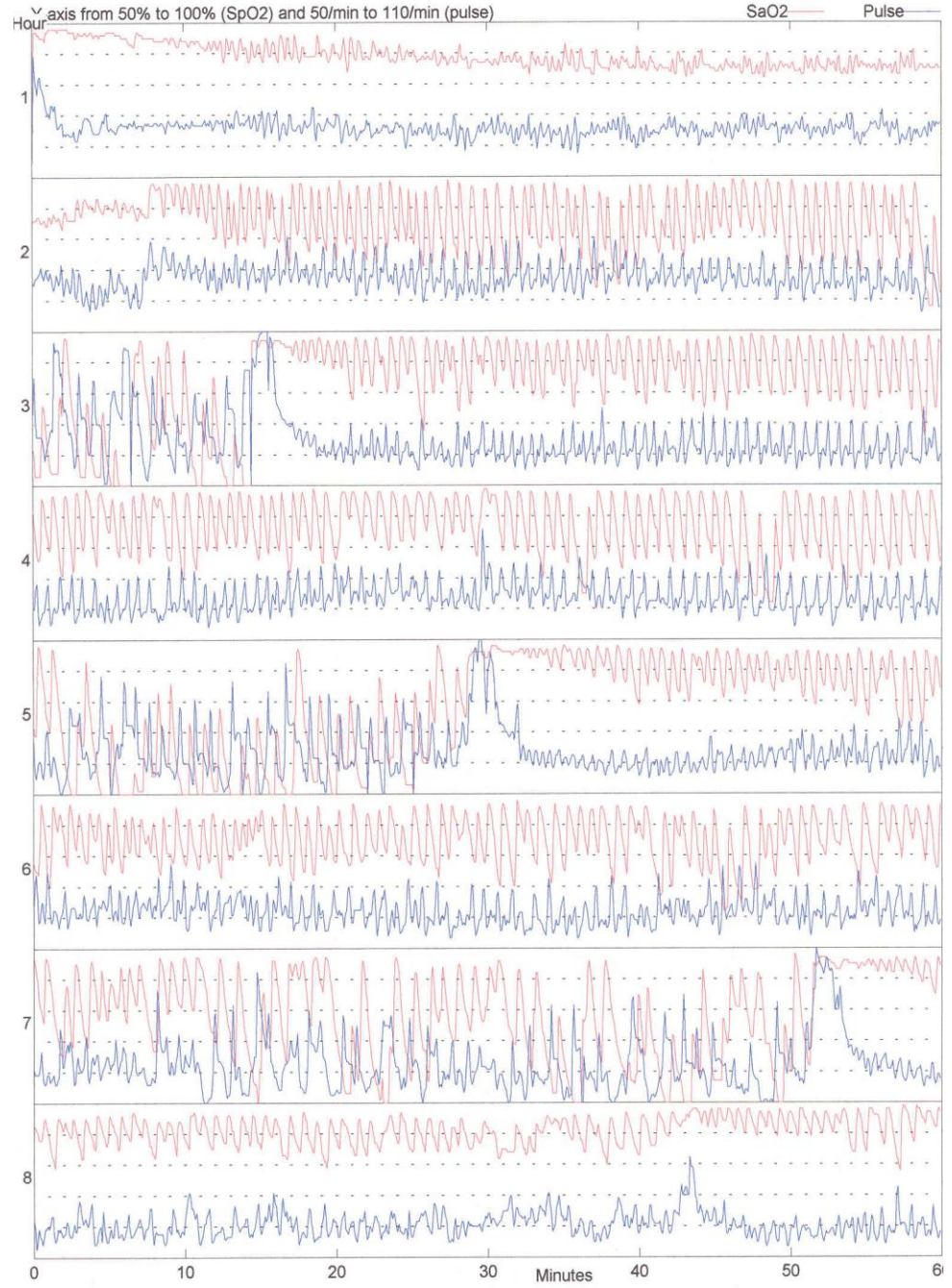
# So how do we diagnose OSAS?







OXIMETRY and PULSE RATE PLOTS for Brian GRIBBEN 23:16:00 13/11/2001



# Treatments for Obstructive Sleep apnoea

- Avoid alcohol/sedatives
- Weight reduction
- Correction of anatomical abnormalities
- Nasal CPAP
- Mandibular advancement splints
- (Adjunctive modafinil)



# Treatments for Obstructive Sleep apnoea

- Avoid alcohol/sedatives
- Weight reduction
- Correction of anatomical abnormalities
- **Nasal CPAP**
- Mandibular advancement splints
- (Adjunctive modafinil)



# Obesity Hypoventilation syndrome

- Watch for those who need nocturnal ventilation not CPAP

# So what do we know and what do we need to know?

- Lots of things can happen to ventilation during sleep
- OSAS is probably common
- OSAS is probably underdiagnosed
- OSAS is associated with an excess risk of cardiac and cerebrovascular disease
- OSAS causes profound daytime sleepiness
- OSAS is associated with a significant risk of Road Traffic Incidents
- OSAS is easy to treat and treatment is cheap and highly cost effective

Scottish Intercollegiate Guidelines Network

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## Management of Obstructive Sleep Apnoea/Hypopnoea Syndrome in Adults

A national clinical guideline

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This guideline is endorsed by the British Thoracic Society

June 2003

[www.sign.ac.uk](http://www.sign.ac.uk)

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## Bronchiectasis:

The word bronchiectasis is derived from the Greek meaning stretching or extension of the air pipes. Nowadays we usually define bronchiectasis as a condition characterised by chronic dilatation of one or more bronchii.

# Causes of Bronchiectasis:

- **Congenital causes and congenital predispositions to bronchiectasis**
  - 1. Kartageners Syndrome (Situs Inversus, Rhinosinusitis and Bronchiectasis)
  - 2. Other types of ciliary dysfunction
  - 3. Cystic Fibrosis
  - 4. Hypogammaglobulinaemia
  - 5. Homozygous alpha 1 antitrypsin deficiency
- **Acquired Bronchiectasis**
  - 1. Childhood pneumonia (whooping cough, measles etc.)
  - 2. Foreign body inhalation
  - 3. Tuberculosis
  - 4. Suppurative pneumonia
  - 5. Bronchopulmonary aspergillosis (which characteristically gives proximal airway bronchiectasis)
  - 6. Bronchial obstruction secondary to adenomas and carcinomas
  - 7. Diseases causing extensive fibrosis, for example, connective tissue disorders such as rheumatoid arthritis
  - 8. Associated with inflammatory bowel disease



## Clinical Features of Bronchiectasis:

- Characteristic symptoms of bronchiectasis are of a chronic cough productive of copious quantities of sputum. This may be complicated by haemoptysis and by pneumonia. Breathlessness may be a feature depending upon the extent of the pulmonary damage.
- Finger clubbing may be present in cases of extensive bronchiectasis and coarse crackles are often audible over areas of bronchiectasis on auscultation

# Investigations for suspected bronchiectasis:

- **Blood tests**

- White cell count
- ESR and C-reactive protein
- Immunoglobulins
- Aspergillus precipitin test
- Rheumatoid factor
- Alpha 1 antitrypsin levels

- **Sputum**

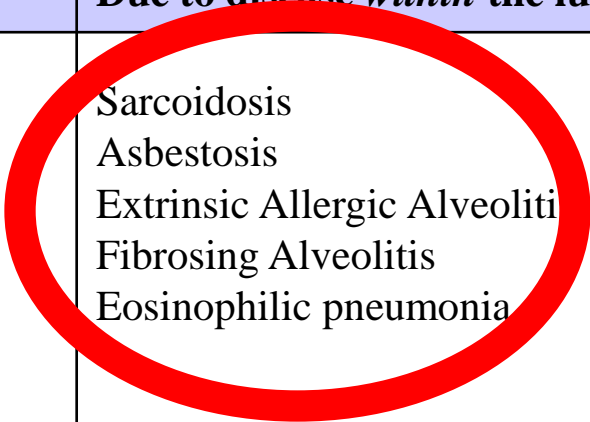
- AFB
- Culture and sensitivity
- Eosinophil count

- **Imaging**

- Chest radiograph
- CT sinus examination
- High resolution thin section CT scan

- **Other investigations**

- Fibreoptic bronchoscopy
- Sweat test
- Aspergillus skin test
- Semen analysis
- Sacharin Test
- Tests of Ciliary Function

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Tuberculosis Infective bronchitis Pneumonia Empyema	Pulmonary emboli Pulmonary hypertension

# Diffuse Parenchymal Lung Disease

- 200 different diseases
- Common feature is a presentation of increasing breathlessness and widespread shadowing on the chest radiograph

# Diffuse parenchymal lung disease

- Acute?
- Episodic?
- Chronic?

# Diffuse parenchymal lung disease

- Acute?
  - Infection
  - Allergy to drugs, fungi or worms
  - Toxins, eg drugs (amiodarone, cytotoxics)
  - Haemodynamic (Heart failure, renal failure)
  - Vasculitic (Goodpastures, Churg Strauss, Wegeners)
  - ARDS
  - Unknown (COP, Eosinophilic pneumonia)
- Episodic?
- Chronic?

# Diffuse parenchymal lung disease

- Acute?
- Episodic?
  - Eosinophilic pneumonia
  - Vasculitis
  - Pulmonary Haemorrhage
  - Churg Strauss
  - Extrinsic Allergic Alveolitis
  - COP
- Chronic?

# Diffuse parenchymal lung disease

- Acute?
- Episodic?
- Chronic?
  - Associated occupation or environment?
  - Associated Drugs or Toxins?
    - Amiodarone, Gold, Bleomycin, Heroin, Radiotherapy
  - Associated Systemic diseases?
  - No evidence systemic disease or external agent



# Diffuse parenchymal lung disease

- Acute?
- Episodic?
- **Chronic?**
  - Associated occupation or environment?
  - Associated Drugs or Toxins?
  - **Associated Systemic diseases?**
  - **SLE, RA, Wegeners, Goodpastures, Sarcoidosis, Ankylosing spondylitis**
  - No evidence systemic disease or external agent

# Diffuse parenchymal lung disease

- Acute?
- Episodic?
- Chronic?
  - Associated occupation or environment?
  - Associated Drugs or Toxins?
  - Associated Systemic diseases?
  - No evidence systemic disease or external agent
  - Cryptogenic fibrosing alveolitis
  - Lymphangiomyomatosis
  - Langerhans Giant Cell Histiocytosis

## What do you need to know?

- ILD can be due to drugs or occupation and can be associated with systemic diseases
- Pulmonary Fibrosis (Fibrosing alveolitis) is common in the Elderly and frequently manifests as a cough or shortness of breath and crackles are audible on auscultation
- A little more knowledge on some specific types of ILD may be helpful, eg Sarcoidosis

# Sarcoidosis:

A granulomatous disorder of unknown aetiology characterised by epithelioid cell tubercles, without caseation, proceeding to either resolution or conversion to avascular hyaline fibrous tissue. May affect any system of the body but commonly presents with bilateral hilar lymphadenopathy or pulmonary infiltrates. (>90% of patients have intrathoracic disease).

# Possible presentations of Sarcoidosis

## Chest Physician

BHL

Pulmonary infiltrates

Pulmonary fibrosis

## Ophthalmologist

Uveitis

Conjunctivitis

Sjogren-like syndrome

## Neurologist

Peripheral neuropathy

Meningitis

Isolated cranial N palsy

S.O.L.

Post-Pituitary lesion

Transverse myelitis

## Rheumatologist

Polyarthralgia

Bone Cysts

## Gastroenterologist

Hepato-splenomegaly

Salivary gland enlargement

## Dermatologist

Erythema Nodosum

Plaques

## Cardiologist

Conduction Disorders

C.C.F. (Myocarditis)

## E.N.T. Surgeon

Nasal Granuloma

Laryngeal plaques

## General Physician

Hypercalcaemia/Hypercalcuria

Renal Disorders

Renal Calculi

Lymphadenopathy

# Investigation of suspected sarcoidosis

Chest Radiograph and E.C.G.

Tuberculin test

Biopsy

Bronchial Mucosal biopsy

Transbronchial lung biopsy

Nasal biopsy

Conjunctival biopsy

Skin Biopsy

Liver biopsy

Lymph node biopsy

Kveim biopsy (Of historical interest only)

Serum and Urinary Ca<sup>+</sup>

Angiotensin Converting Enzyme

Broncho-alveolar lavage

Gallium Scanning

# Staging of Intrathoracic Sarcoidosis

<u>Stage</u>	<u>Chest Radiograph</u>	<u>% of Patients at Presentation</u>
0	Normal	8%
I	Hilar Adenopathy	51%
2	BHL plus infiltrates	
3	Pulmonary Infiltrates	29%
4	Pulmonary Fibrosis	12%

# Sarcoidosis - Possible treatments

- 1) Time (Spontaneous resolution)
- 2) Topical and Systemic Steroids
- 3) Azathioprine plus steroids
- 4) Hydroxychloroquine
- 5) Methotrexate
- 6) Cyclosporin A
- 7) Pentoxifylline
- 8) Transplantation

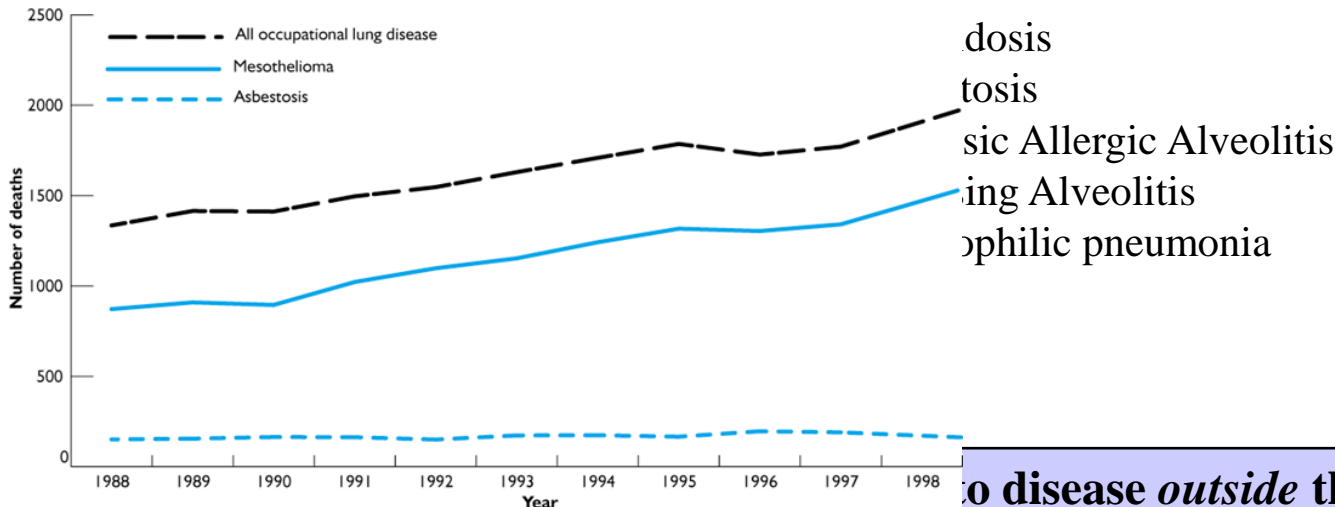


# Airways diseases

# Small lung disorders

(also known as "restrictive disorders")

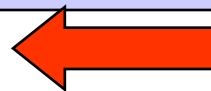
## to disease *within* the lungs



## to disease *outside* the lung

Asthma  
C.O.P.D.  
Bronchiectasis  
Cystic Fibrosis  
Obliterative Bronchiolitis

Mesothelioma  
Pneumothorax  
Scoliosis  
Respiratory muscle weakness



Deaths due to asbestos related lung disease increasing significantly

## Infections

## Pulmonary vascular disorders

Tuberculosis  
Infective bronchitis  
Pneumonia  
Empyema

Pulmonary emboli  
Pulmonary hypertension

# Exposure to asbestos may lead to:

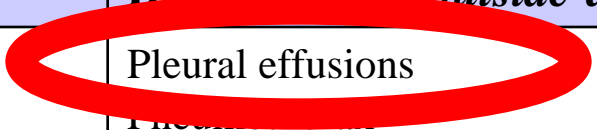
- Asbestosis
- Mesothelioma
- Benign Pleural plaques
- Diffuse pleural thickening

# Occupations involving exposure to asbestos:

- Asbestos industry (mining, manufacture)
- Naval dockyards
- Dock workers in general
- Builders, ladders, plumbers, demolition workers, electricians, boilerhousemen
- Brake lining manufacturers
- etc

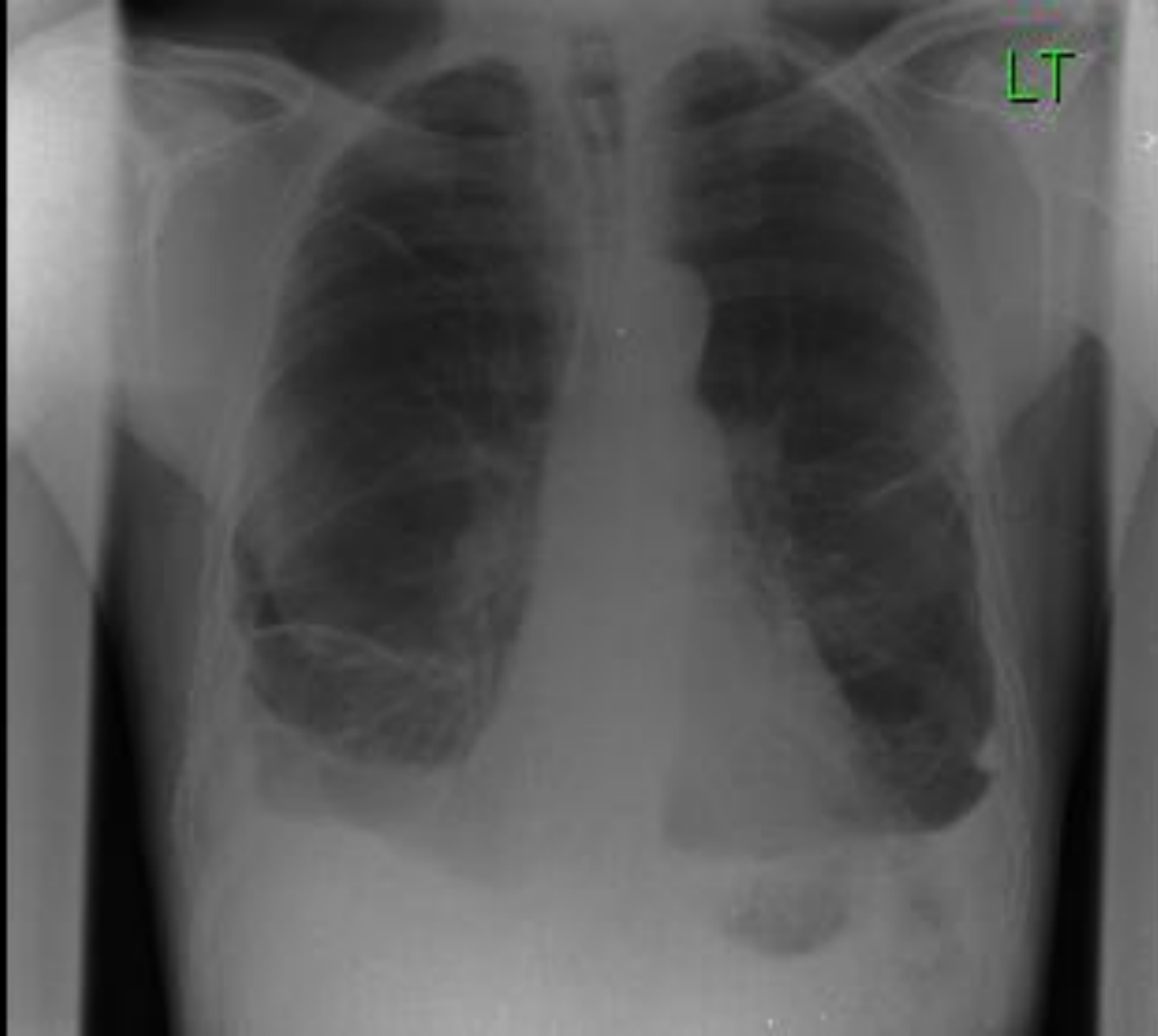
*and the wives and family of the same*

<b>Airways diseases</b>	<b>Small lung disorders</b> (also known as “restrictive disorders”)
<b><i>Localised obstruction</i></b>	<b>Due to disease <i>within</i> the lungs</b>
<ul style="list-style-type: none"> <li>Sleep apnoea</li> <li>Laryngeal carcinoma</li> <li>Thyroid enlargement</li> <li>Vocal cord dysfunction</li> <li>Relapsing Polychondritis</li> <li>Tumours</li> <li>Post tracheostomy stenosis</li> <li>Foreign bodies</li> <li>Bronchopulmonary dysplasia</li> </ul>	<ul style="list-style-type: none"> <li>Sarcoidosis</li> <li>Asbestosis</li> <li>Extrinsic Allergic Alveolitis</li> <li>Fibrosing Alveolitis</li> <li>Eosinophilic pneumonia</li> </ul>
<b><i>Generalised obstruction</i></b>	<b>Due to disease <i>outside</i> the lung</b>
<ul style="list-style-type: none"> <li>Asthma</li> <li>C.O.P.D.</li> <li>Bronchiectasis</li> <li>Cystic Fibrosis</li> <li>Obliterative Bronchiolitis</li> </ul>	<ul style="list-style-type: none"> <li>Pleural effusions</li> <li>Pneumothorax</li> <li>Scoliosis</li> <li>Respiratory muscle weakness</li> <li>Obesity</li> </ul>



<b>Infections</b>	<b>Pulmonary vascular disorders</b>
<ul style="list-style-type: none"> <li>Tuberculosis</li> <li>Infective bronchitis</li> <li>Pneumonia</li> <li>Empyema</li> </ul>	<ul style="list-style-type: none"> <li>Pulmonary emboli</li> <li>Pulmonary hypertension</li> </ul>

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## **Box 2 Causes of transudative pleural effusions**

### **Very common causes**

- Left ventricular failure
- Liver cirrhosis
- Hypoalbuminaemia
- Peritoneal dialysis

### **Less common causes**

- Hypothyroidism
- Nephrotic syndrome
- Mitral stenosis
- Pulmonary embolism

### **Rare causes**

- Constrictive pericarditis
- Urinothorax
- Superior vena cava obstruction
- Ovarian hyperstimulation
- Meigs' syndrome

## **Box 3 Causes of exudative pleural effusions**

### **Common causes**

- Malignancy
- Parapneumonic effusions

### **Less common causes**

- Pulmonary infarction
- Rheumatoid arthritis
- Autoimmune diseases
- Benign asbestos effusion
- Pancreatitis
- Post-myocardial infarction syndrome

### **Rare causes**

- Yellow nail syndrome
- Drugs (see box 1)
- Fungal infections

Two Final points!



# Orthopnoea

- Most causes of breathlessness are associated with worsening on lying flat
- Orthopnoea is classically a symptom of cardiac failure but beware of diaphragm weakness as a cause
- The amyotrophic variant of motor neurone disease frequently presents with orthopnoea (as may Guillain Barre)

# Diffuse parenchymal lung disease in the elderly

Crackles on auscultation and no response to diuretics and no other signs of heart failure ?

**THINK** ? Fibrosing alveolitis

Check spirometry

Check Chest Radiograph

# Diffuse parenchymal lung disease in the elderly

Crackles on auscultation and no response to diuretics and no other signs of heart failure ?

**For exam purposes: THINK**

If crackles are fine, basal and bilateral the likeliest diagnosis is diffuse parenchymal lung disease

If coarse, especially if localised, more likely to reflect bronchiectasis

## Further Information:



**Respiratory Guidelines**

**[www.brit-thoracic.org.uk](http://www.brit-thoracic.org.uk)**

