Getting the Basics Right

Recognising and diagnosing rarer lung conditions

A summary of PCRS-UK opinion sheets input from **Dr Iain Small** and **Dr Basil Penney**

As a primary care organisation, the first priority for PCRS-UK is trying to get the diagnosis right for people with asthma and COPD because these are the patients that we see and manage most commonly. Nevertheless, there is a need to be able to recognise and appropriately refer and manage patients who may have rarer lung conditions.

Rarer respiratory conditions are, by definition, uncommon and each practice will have only a handful of such patients. The most important things that clinicians need to know are:

- Limits of your own knowledge
- How such patients present
- Referral pathways to secondary care
- What the local Community Respiratory Team can provide

However, these patients will continue to be looked after in primary care and so clinicians will also need to know:

- Where to get information for a patient and themselves when a diagnosis is made
- The typical natural history of the disease
- What to do in the face of an 'exacerbation' of symptoms
- Rarer conditions may co-exist in patients with commoner respiratory disease. Make sure you treat the treatable!

Acknowledgements and further information
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Bronchiectasis

In children this is commonly caused by cystic fibrosis (CF): patients presenting as adults have a number of different causes. Presenting features typically include chronic productive cough, dyspnoea and wheeze. There are no good up-to-date figures for prevalence but, as a guide, the average full-time GP with a list size of about 2,000 patients would expect to manage approximately two cases of bronchiectasis.

- High-resolution computerised tomography (HRCT) is the investigation of choice for diagnosis.
- Common causes of bronchiectasis include CF, foreign bodies and aspiration, immune deficiency, asthma associated with allergic bronchopulmonary aspergillosis and connective tissue disease.
- Asthma is considered to be the cause of bronchiectasis when no other cause is found, therefore be prepared to look for and manage asthma in these patients.
- All patients should be taught airway clearance techniques by a physiotherapist and pulmonary rehabilitation should be offered to those troubled by dyspnoea.
- Exacerbations present as worsening of cough, sputum or dyspnoea. Treat with amoxicillin or clarithromycin, having first sent a sputum sample. Where possible, GPs should treat according to sensitivity.
- Chronic bacterial colonisation of the airways may require long-term antibiotics.
- Prognosis is generally good in non-CF patients although general health can be poor, especially in those whose lungs are colonised with Pseudomonas where progressive deterioration may occur.

Interstitial Lung Disease

The term encompasses a number of conditions that result in parenchymal lung disease which can present as:

- shortness of breath
- cough
- fine inspiratory crackles
- restrictive spirometry
- (possibly) abnormal chest X-ray findings.

Although surgical biopsy is the gold standard for diagnosis, HRCT and pulmonary function tests will often provide sufficient diagnostic certainty.

The commonest type of interstitial lung disease (ILD), formerly termed cryptogenic fibrosing alveolitis, has around 2,000 new cases each year in England and Wales with an average survival of three years from diagnosis. Currently, no treatment is proven to affect progression. This group is now recognised to be made up of two distinct forms: idiopathic pulmonary fibrosis (IPF) with a 5-year survival of 10–15% and non-specific interstitial pneumonia with a better prognosis of >50% survival over 5 years. Other forms include those associated with connective tissue disorders (approximately 10% of the total), hypersensitivity (bird exposure and drug-related being most common in the UK) and sarcoidosis (see below).

Suspected cases should be referred to the local chest physicians.

- Inhalers: bronchodilators and corticosteroids are of no proven value.
- Oral steroids have no evidence-based place in the treatment of acutely worsening symptoms in the community.
- Haemoptysis is rarely associated with ILD alone. Always investigate for cancer, pneumonia and pulmonary embolus.
- Treatment for the disease is often 'Best Supportive Care' and may include referral to palliative care services.
- Despite symptoms of breathlessness, oxygen has little place unless the patient is chronically hypoxic, although a literature review did find some weak evidence for a small benefit from ambulatory oxygen.

There are now two licensed treatments for ILD: pirfenidone and the more recent monoclonal antibody, rituximab. Each has been approved by NICE and the Scottish Medicines Consortium (SMC) for use under strict restrictions of lung function, diagnostic testing (characteristic pattern on HRCT) and a monitored response to treatment. The role of primary care is to identify patients with suspicious symptoms and signs and a restrictive pattern on spirometry and refer promptly. Neither drug may currently be used unless the FVC is below 80% predicted, although this may change in time once more accurate diagnostic assessment identifies the early responding group.

Lung Cancer

Lung cancer is one of the most common cancers in the UK but, although a common cancer, most GPs will see only 1–2 new cases a year.

Lung cancer is linked to smoking in over 85% of cases. The relative risk for someone who smokes 20 cigarettes per day is approximately 20 times greater than that of a lifetime non-smoker. Lung cancer death risk is around 24 times higher in smokers of 25+ cigarettes per day and 39 times higher in smokers of 42+ cigarettes per day. Female smokers appear more susceptible. Stopping smoking before middle age avoids 90% of the risk. Passive smoking increases risk. Patients with COPD are 3–5 times more likely to develop lung cancer than smokers without COPD. Other risk factors include occupational exposure to asbestos, nickel, cadmium and arsenic.

The symptoms of lung cancer are similar to other more common diseases and making a diagnosis is often not easy. Symptoms such as haemoptysis tend to lead to rapid referral, but more non-specific symptoms of fatigue and weight loss may not always alert the clinician to the possibility of lung cancer. Perhaps the most common presentation is cough that does not improve after a few weeks.

Summary of NICE guidance on referral for suspected lung cancer

Urgent referral for CXR with symptoms

- Haemoptysis or unexplained or persistent, more than 2 weeks

- Cough
- Dyspnoea
- Chest/shoulder pain
- Weight loss
- Chest signs
- Hoarseness
- Finger clubbing
- Supraclavicular or cervical persistent lymphadenopathy
- Features of metastases: brain, bone, liver, skin

Urgent referral to a member of a lung cancer multidisciplinary team (MDT) – chest physician

- If CXR suggests lung cancer.
- Even if CXR is normal, there may be high suspicion of lung cancer clinically.
- Persistent haemoptysis in smoker or ex-smoker over the age of 49 years.
- Signs of SVC obstruction or stridor in younger patients.

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A CXR will show abnormalities in over 90% of patients with lung cancer, but it is important to remember that a normal CXR does not exclude lung cancer if there is a high index of clinical suspicion.

Some areas now have direct access to CT imaging if the above clinical criteria are met. It is worth finding out if you work in one of these areas.

Referral

There are well established referral pathways to the lung cancer MDT for patients to be seen within two weeks. Investigation, staging and diagnosis should be completed within 31 days and specialist treatment begun within 62 days of GP referral.

The primary care role post-diagnosis is important and includes:

- General advice and education about the disease and its treatments.
- Counselling and support for patient and carers.
- Regular review, assessing symptoms, nutrition, mood and social situation.
- Palliative care support within the MDT.

Tuberculosis (TB)

Symptoms:

- Persistent cough (>3 weeks)
- Fevers
- Night sweats
  - Weight loss
  - Lethargy
  - Loss of appetite
  - Chest pain
  - Haemoptysis

Diagnosis is usually achieved with chest X-ray and three spontaneously produced sputum samples. Suspicion of pulmonary TB warrants rapid access to a chest physician with expertise in the management of TB.

Every patient being treated for TB should have a named key worker responsible for education and ensuring adherence with treatment.

Treatment usually consists of six months anti-tuberculous drugs, initially rifampicin, isoniazid and either pyrazinamide or ethambutol for two months and then a further four of rifampicin and isoniazid. This is considered curative and patients are not generally offered follow-up. Poor adherence to treatment encourages drug resistance.

Sputum microscopy positive patients are usually considered to be infectious until they have completed two weeks of treatment.

Latent TB is said to be present in those individuals with a strongly positive skin (Mantoux or Heaf) test. They have a 10–15% risk of developing active disease, especially in association with other risk factors that reduce immune function.

BCG vaccination is now offered on an 'at risk' basis rather than to all schoolchildren. There is little benefit of protection from this vaccine in those over 35 years.

Contact tracing is done by key workers and usually aimed at close household contacts, except in outbreaks involving institutions (schools, hospitals, prisons) where the Health Protection Agency or similar will set up specific programmes.

Non-tuberculous Mycobacterium Infections

These opportunistic infections most commonly affect middle aged and older men with COPD (typically emphysema) and previous TB infections. They present with TB-like symptoms and should be looked after as hospital outpatients.

If they require treatment this often extends to two years and is determined by the sensitivities of the organism.

Aspergillosis

This term covers a large number of diseases that involve infection and growth of this fungus as well as allergic responses. The commonest presentations are in the lung and include:

- Invasive pulmonary aspergillosis – generally occurs in immunocompromised patients.
- Allergic bronchopulmonary aspergillosis – affecting patients with asthma, bronchiectasis and cystic fibrosis; treated with long-term steroids.
- Chronic pulmonary aspergilloma – long-term aspergillus infection usually in those with underlying lung disease.
- Severe asthma with fungal sensitisation (SAFS) – one of the causes of difficult to control asthma.

Sarcoidosis

This is a multisystem disease of unknown cause predominantly affecting young and middle aged patients, most often affecting the lungs (>90% of cases) or skin. Spontaneous remission occurs in up to 60% within six months, particularly in milder forms. In spite of this, the course and prognosis are variable and difficult to predict; however, systemic treatment is often unnecessary. Common respiratory symptoms include:

- non-productive cough
- breathlessness
- wheeze

Other common non-respiratory symptoms include fatigue, uveitis (25%), erythema nodosum (25%); 25–50% will have an inflammatory arthritis most commonly involving the ankle, knee, wrist or elbow. Hypercalcaemia can cause nephrocalcinosis, which causes renal impairment and is a major cause of chronic renal failure. Hypercalcaemia is one of the indications for oral glucocorticoid treatment along with ocular sarcoid not responding to topical treatment and neurological and cardiac manifestations.

Cardiac sarcoid is rare but life-threatening and symptoms of palpitations and syncope should be taken seriously. Investigations include spirometry (typically showing a restrictive defect) and chest X-ray (showing hilar lymphadenopathy).

Serum angiotensin converting enzyme (ACE) is often raised but has a limited use in diagnosing or monitoring the disease as it is relatively non-specific.

Pulmonary Hypertension

This group of conditions, characterised by a raised mean pulmonary artery pressure, has a prevalence of approximately 25 treated patients per million. It presents in a very non-specific manner and should be suspected in anybody presenting with breathlessness without overt signs of cardiorespiratory disease.

There is no definitive test in primary care although chest X-ray, ECG and echocardiography can all be suggestive, particularly the latter.

There is frequently a diagnostic delay of three years between the first presenting symptom and diagnosis. Patients are largely looked after in specialist centres but GPs need to be aware that their symptoms can break unpredictably through drug treatment.

Treatment often involves using drugs or combinations of drugs that require monitoring similar to that for disease-modifying anti-rheumatic drugs (DMARDs) which clinicians in primary care may be asked to perform.

Chronic thromboembolic pulmonary hypertension follows 12–24 months after a pulmonary embolus in about 4% of patients and presents with continued or recurrent breathlessness. It is important because it is treatable by endarterectomy.