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Opinion

What every GP should know about rarer respiratory conditions

What every GP should know

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

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

The British Thoracic Society has produced useful guidelines on referral to specialist services that includes standards for shared care. (available from http://www.brit-thoracic.org.uk (guideline section))

High Resolution CT Scanning. (HRCT)

This is a useful radiographic technique that provides detailed images of the lung. Thin sections (1-2mm) are viewed at 10-40mm intervals giving a representative view of the lungs though only covering about 10% of the total. For this reason they are important for the diagnosis of conditions such as bronchiectasis and pulmonary fibrosis but unsuitable for assessment of lung cancer and other localised disease where a standard contrast CT scan of the chest (and abdomen) is necessary. As referral pathways develop and more of the diagnostic work-up is done in primary care this may become a modality more commonly requested by GPs.

Interstitial Lung Disease

The term encompasses a number of conditions that result in parenchymal lung disease which can present to GPs 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, formerly termed Cryptogenic Fibrosing Alveolitis, has around 2,000 new cases each year in England and Wales with an average survival of 3 years from diagnosis and no treatment currently 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 (approx 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 recent literature review found some weak evidence for a small benefit from ambulatory oxygen[1]

Sarcoidosis

This is a multisystem disease of unknown cause predominantly affecting young and middle aged patients most often affecting the lungs (>90% cases) or skin. Spontaneous remission commonly occurs (up to 60%) within 6 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 ankle, knee, wrist or elbow. **Hypercalcaemia** can cause nephrocalcinosis, which accounts for half of sarcoid patients with renal impairment and is the 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 required 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 Vasculitis

Systemic vaculitides such as Wegners Granulomatosis, Churg Strauss Syndrome and Polyarteritis Nodosa can all affect the respiratory tract (upper and lower) as well as other organ systems, commonly involving a degree of renal damage. They can present with breathlessness, cough (dry or productive) and haemoptysis. Nearly three-quarters of patients present initially with symptoms referable to the ears, nose and throat. X-Rays can show nodules, pulmonary infiltrates and pleural effusions. Serum anti-cytoplasmic neutrophil antibody (ANCA) levels are almost invariably raised.

Investigation and treatment will normally take place in secondary care and management is often multi-speciality.

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 each GP will have a few such patients.

- 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 for 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.
- 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.

British Lung Foundation (BLF)
The BLF is a charity dedicated to supporting people with lung disease as well as funding research into respiratory conditions. Their website (www.lunguk.org) gives contact details of the Breathe Easy patient groups that campaign and provide a local network and regular meetings for those with many lung disorders. There are also a number of patient information leaflets regularly updated and available to view and print on the site.

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 cardio-respiratory 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 3 years between 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.

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.

Tuberculosis (TB)

- Making a diagnosis of TB

  Consider it!

  Symptoms:
  - Persistent cough (> 3 weeks)
  - Fevers
  - Night sweats
    - o Weight loss
    - o Lethargy
    - o Loss of appetite
    - o Chest Pain
    - o 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 management of TB
- Every patient being treated for TB should have 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 2 weeks of treatment.

Latent TB is said to be present in those individuals with a strongly positive skin (Mantoux or Heaf) test and 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 immune-compromised patients.
- Allergic bronchopulmonary aspergillosis – affecting patients with asthma, bronchiectasis and cystic fibrosis; treated with long term steroids.
- Chronic pulmonary and aspergilloma – long term aspergillus infection usually in those with underlying lung disease.
- Severe asthma with fungal sensitisation – one of the causes of difficult to control asthma.

Reference

1. Bajwah S, *et al. Thorax* 2013; 68:867–879. doi:10.1136/thoraxjnl-2012-202040) http://dx.doi.org/10.4104/pcrj.2013.00100

Further reading and resources:

Condition	Guideline or resource	Available from
General resources	BTS statement on criteria for specialist referral, admission, discharge and follow-up for adults with respiratory disease	British Thoracic Society http://www.brit-thoracic.org.uk/Clinical-Information
	Patient information on a range of lung conditions	British Lung Foundation http://www.lunguk.org
	PCRS-UK Opinion No.36: The Differential Diagnosis of the Breathless Patient	PCRS-UK http://www.pcrs-uk.org/pubs/opinionsheets.php
	Primary Care Respiratory Journal Case Study: Kaplan A, Gruffydd-Jones K, van Gemert F, Kirenga BJ, Medford ARL. A woman with breathlessness: a practical approach to diagnosis and management. Prim Care Respir J 2013;22(4):468-476.	http://www.thepcrj.org/journ/citation_export.php?type=part&article_id=1079 DOI: http://dx.doi.org/10.4104/pcrj.2013.00100
Interstitial Lung Disease	BTS Interstitial lung disease guideline *Thorax* 2008;**63**(Suppl):v1-v58	http://www.brit-thoracic.org.uk/clinical-information/interstitial-lung-disease-(dpld).aspx
Sarcoidosis	Dempsey OJ, Paterson EW, Kerr KM, Denison AR. Sarcoidosis. *BMJ* 2009;**339**:doi:10.1136/bmj.b3206	http://www.bmj.com/content/339/bmj.b3206.full?sid=2ba0076d-116b-497c-bdca-7f93c7f3ad66
Bronchiectasis	Guideline for non-CF Bronchiectasis *Thorax* 2010;**65**(Suppl):i1-i58	http://www.brit-thoracic.org.uk/clinical-information/bronchiectasis.aspx
	ten Hacken NHT, Wickstra PJ, Kerstjens HAM. Treatment of bronchiectasis in adults *BMJ* 2007;**335**:1089-1093	http://www.bmj.com/content/335/7629/1089.full
Tuberculosis	Tuberculosis: NICE Guidance	http://guidance.nice.org.uk/CG33/NICEGuidance/pdf/English
Non-tuberculous Mycobacterium Infection	Subcommittee of the Joint Tuberculosis Committee of the British Thoracic Society. Management of Opportunist Mycobacterial Infections: Joint Tuberculosis Committee Guidelines 1999. *Thorax* 2000;**55**:210-218	http://www.brit-thoracic.org.uk/clinical-information/opportunist-mycobacteria.aspx
Pulmonary hypertension	Galie N, Torbicki A, Barst R, *et al* for the Task Force on Diagnosis and Treatment of Pulmonary Arterial Hypertension of the European Society of Cardiology. Guidelines on diagnosis and treatment of pulmonary arterial hypertension. *Eur Heart J* 2004;**25**:2243–78	http://www.escardio.org/guidelines-surveys/esc-guidelines/Pages/pulmonary-arterial-hypertension.aspx
Pulmonary vasculitis	Burns A. Pulmonary Vasculitis. *Thorax* 1998;**53**:220-227	http://thorax.bmj.com/content/53/3/220.full
Aspergillosis	A comprehensive resource providing a wide range of information about the fungus Aspergillus and the diseases it can cause.	The Aspergillus Website http://www.aspergillus.org.uk

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