

ASBESTOS RELATED LUNG DISEASE

Version 2 Final

Document control

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Changes since last version

Description

Asbestos is the generic term for a group of fibrous minerals.

Asbestos fibres are highly resistant to physical and chemical breakdown. This gives them their commercial value in applications such as fireproofing, insulation, reinforced cement, and brake linings. In 1970, a voluntary ban on the use of asbestos was introduced in the UK, and it was given legislative force in 1986.

Asbestos was mined on a large scale in Canada, South Africa, and the former Soviet Union. The asbestos-bearing rock was crushed to release the fibres, which were then processed, packed into impermeable bags and transported to the manufacturer.

The main types of asbestos fibres are:

- **White** (Chrysotile).
Flexible and easily inhaled.
- **Blue** (Crocidolite).
Stiff, straight and able to travel deep into the lung.
Very strongly associated with the development of mesothelioma.
The most dangerous form of asbestos.
- **Brown** (Amosite).
Stiff, straight and able to travel deep into the lung.
Associated with the development of mesothelioma.

Inhalation of asbestos fibres can cause Asbestos Related Lung Disease:

- a) **Asbestosis**
- b) **Pleural Plaque**
- c) **Diffuse Pleural Thickening**
- d) **Mesothelioma.**

Asbestos related lung disease is increasing in prevalence, and is not expected to peak until 2020.^[1]

It is likely to become an increasingly common aspect of disability analysis medicine.

Aetiology

Blue and brown asbestos fibres are thinner than 3µm and longer than 10µm. (A human hair is 100µm in diameter.) This makes the fibres narrow enough to be inhaled into the alveolar part of the lung, but too long to be removed by macrophages. Asbestos fibres can persist in the body for decades.

Fibrosis or 'scarring' of the lungs is caused by the body's inflammatory response to the asbestos.

In non-malignant asbestos related lung disease, the prevalence and severity of the conditions increase with increasing exposure to asbestos.^{[2][3]}

Asbestosis

Asbestosis which is a symptom complex rather than a specific disease results in interstitial pulmonary fibrosis caused by the inhalation of asbestos.

It was originally described in the 1900s and the importance of its occupational cause was recognised by epidemiological studies in the 1930s. As early as the first century AD, Pliny recorded that the weavers of asbestos wicks for the lamps of the vestal virgins wore masks for respiratory protection.

Asbestosis occurs in those who have had regular exposure.^[2]

However, women, whose only contact with asbestos has been washing their husband's clothes, have developed asbestos related lung disease.^[4]

Pleural Plaque

This is an area of local fibrous thickening on the lung pleura.

The likelihood of developing pleural plaques increases with increased exposure to inhaled asbestos. They generally develop 10 to 20 years after exposure.

Pleural plaque is usually asymptomatic, and often presents as an incidental finding on a CXR.^[4]

Diffuse Pleural Thickening

This is uniform smooth thickening of the pleura.

Pleural effusions cause inflammation and fibrosis. Recurrent episodes lead to pleural thickening.

Diffuse pleural thickening is like a 'straight jacket' around the lungs. The more it spreads, the more it limits lung expansion.

Benign Mesothelioma

Benign fibrous mesothelioma is a rare tumour. It is neither associated with exposure to asbestos nor related to the development of malignant mesothelioma. It will not be discussed further in this protocol.

Malignant Mesothelioma

Mesothelioma is a malignant tumour of mesothelial cells derived from the pleura (most common), pericardium (rare) and peritoneum (rare.) The tumour begins as a local mass, often associated with a pleural effusion. It gradually spreads to encase the lungs and extends to involve the chest wall and the pericardium. However, clinical problems associated with metastatic spread are uncommon.

Prior exposure to asbestos is a prerequisite for the diagnosis of mesothelioma as a prescribed disease PD D3.

In heavily exposed individuals, the lifetime risk of developing mesothelioma is approximately 10%. Individuals who have never been exposed to asbestos have a lifetime risk of only one in a million. However, there does not seem to be a simple relationship between the amount of exposure and the risk of developing mesothelioma.^[4]

Exposure to blue (crocidolite) asbestos poses the greatest risk.

The latent period after exposure is between 15 and 40 years, so mesothelioma usually presents between the ages of 50 and 70.^[4]

Mesothelioma affects five men for every affected woman. This reflects traditional patterns of employment.^[4]

Epidemiology

The prevalence of asbestos related lung disease is increasing and is expected to peak around 2020.^{[1][5]}

Mesothelioma

- Mesothelioma caused 1527 deaths in the UK during 1998.^[5]
- 1032 new cases of mesothelioma were confirmed in the UK during 1999.^[6]
- Due to the natural history of the disease, (it has an exceptionally long latent period), the death rate from mesothelioma is expected to increase to 3300 per annum by 2020.^[1]
- All occupational lung disease has risen rapidly since the late 1980s due to a 75% increase in the number of mesothelioma deaths.^[5]
- Men born in the 1940s are the worst affected cohort. Malignant mesothelioma may account for about 1% of all their deaths.^[4]

Non-Malignant Asbestos Related Lung Disease

- Benign asbestos related lung disease was the largest cause of new cases of occupational respiratory disease in the UK during 1999. (1256 new cases, 28% of the total.)^[6]
- From 100 - 150 new cases of asbestosis are confirmed in the UK each year.^[4]
- Pleural plaques are the most frequent manifestation of asbestos related lung disease.^[2]

Diagnosis

The Occupational History is crucial in reaching a diagnosis.

Asbestos exposure typically affects people who have worked in the following industries:

- a) Construction and Building
- b) Ship Building (especially pipe-laggers and plumbers)
- c) Dock Workers
- d) Demolition
- e) Asbestos Production
- f) Rail Workers.

Evidence from the occupations recorded on Death Certificates suggests that building workers, especially plumbers, gas fitters, carpenters and electricians are the largest high-risk group.

Asbestosis

Symptoms

The early stages of asbestosis are asymptomatic. When symptoms develop, patients complain of a non-productive cough, and **breathlessness on sustained exertion**.

Examination

The Key Finding On Examination:

Fine end-inspiratory crackles, which persist or increase after coughing.

Finger clubbing is a late, non-specific, feature of asbestosis. Cyanosis is often present in the latter stages of asbestosis and is usually attributable to concomitant COPD. Chest expansion is usually reduced. (Patients often complain that they have difficulty taking a deep breath.)

Investigations

Pulmonary Function Tests

In the early stages of asbestosis, lung function is normal.

As asbestosis progresses, pulmonary function tests reveal a 'restrictive' impairment of lung function. (The FVC and FEV₁ are reduced, but the ratio of FEV₁/FVC is normal or increased.)

Medical Services

Gas Transfer Factor is reduced.

Definitions:

- FVC is the Forced Vital Capacity.
- FEV₁ is Forced Expiratory Volume in 1 second.
- Gas Transfer Factor measures the transfer of inspired carbon monoxide into the pulmonary circulation, and thus indicates the efficiency of oxygen transfer in the lungs.
- Pulmonary function tests are useful for screening and for monitoring progression.

Radiology^[7]

CXR is frequently normal in the early stages of the disease.

In the late stages of extensive asbestosis, the CXR may show a typical basal 'honeycombed appearance.'

High resolution CT scanning is the investigation of choice.

Management

After diagnosis, cases of asbestosis usually remain under the care of a Consultant Respiratory Physician, and are subject to annual review in the chest clinic until it is clear that the condition is stable. Deterioration in the condition should prompt a re-referral to the clinic.

There is no specific treatment for asbestosis but treatment will be that appropriate to the condition it may trigger.

Pleural Plaque

Symptoms

This condition is usually asymptomatic. (If plaques become very large and widespread, symptoms of breathlessness and cough may develop, and the clinical presentation will start to resemble the early stages of diffuse pleural thickening.)

Examination

Usually there are no specific clinical findings on examination. In extensive disease, reduced chest expansion and quiet breath sounds may be found.

Medical Services

Investigations

CXR most often shows multiple bilateral pleural plaques.^[7] (The plaques may be calcified.)

Management

Once the initial diagnosis has been made, routine clinic review is not normally required.

Diffuse Pleural Thickening

Symptoms

Diffuse pleural thickening usually presents following recurrent episodes of pleuritic pain.
Cough and breathlessness on exertion are accompanying features.

Examination

Chest expansion is reduced, and the breath sounds are quiet.

Investigations

As the disease progresses, pulmonary function tests reveal a 'restrictive' impairment of lung function.
Impairment of the Gas Transfer Factor correlates with the degree of pleural thickening.

Management

After diagnosis, cases of diffuse pleural thickening usually remain under the care of a Consultant Respiratory Physician, and are subject to regular review in the chest clinic until it is clear that the condition is stable. Deterioration in the condition should prompt a re-referral to the clinic.

Mesothelioma

Symptoms

Mesothelioma typically presents with chest pain, shortness of breath, day and night sweats and weight loss. (Patients will sometimes describe a persistent flu-like illness.)

Medical Services

Examination

Finger clubbing is rare. Cyanosis is not a feature unless end stage respiratory failure is present.

Tumour invasion commonly occurs at the biopsy site. In advanced disease, tumour deposits may be visible or palpable in the chest wall.

Chest expansion becomes increasingly reduced as the condition progresses.

Stony dullness on percussion and reduced air entry indicates the presence of a pleural effusion.

Investigations

CXR may show features suspicious of mesothelioma. Pleural effusion is a common finding. CT scanning is the radiographic investigation of choice.

A pleural biopsy is obtained for histology.

If necessary, this is achieved by an open procedure with X-ray guidance.

Immunohistochemistry and electron microscopic techniques help to distinguish mesothelioma from other tumours, such as adenocarcinoma.^[8]

CT and MRI scanning are also useful for determining tumour size and resectability.^[8]

Management

Responsibility for the management of a case of mesothelioma is likely to be shared between a Respiratory Physician, a Thoracic Surgeon, an Oncologist and the Primary Care Team. Their roles will depend on the clinical stage of the cancer and the patient's prognosis. As the disease progresses management of these patients is likely to be shared with a palliative care physician.

Differential Diagnosis of Asbestos Related Lung Disease

The presence of pleural plaques is an indication of previous asbestos exposure.

Occupational exposure to airborne pollutants and mineral dusts (for example, asbestos and coal,) is also associated with the development of COPD and interstitial lung disease.^{[9][9]}

- a) COPD. (**See Protocol 'COPD'.**)
- b) Asthma. (**See Protocol 'Asthma'.**)
- c) Other causes of Pleuritic Chest Pain: (e.g. Infection and Pulmonary Embolus.)
- d) Other causes of Pulmonary Fibrosis: (**See Protocol 'Interstitial Lung Disease.'**)

Prevention and Treatment

Prevention

The prevention of asbestos related lung disease relies on the reduction of exposure to the fibres. Workers in contact with asbestos are now required to wear protective clothing and breathe through sophisticated filters. Industry has had to replace asbestos with alternative materials.

Regular medical and radiological examination of workers who use or handle asbestos is a legal requirement. The Health and Safety Executive is responsible for strict regulation of the industry. There is some evidence that removal of the worker from exposure at an early stage is associated with slower progression of asbestos related lung disease.^[2]

It is especially important to avoid smoking because the interaction between smoking and asbestos increases the risk of developing lung cancer.^[10]

In the UK, reports of new cases of occupational respiratory disease by Respiratory and Occupational Physicians are collated by the SWORD and OPRA surveillance schemes. These provide information about the incidence of asbestos related lung disease for research and for the development of protective regulations and legislation.^[6]

Mesothelioma, asbestosis, diffuse pleural thickening and primary carcinoma of the lung (where there is evidence of either asbestosis or bilateral diffuse pleural thickening) are Prescribed Diseases under the Industrial Injuries Provisions of the Social Security Contributions and Benefits Act 1982.

- About 100 people with asbestosis are awarded Industrial Injury Scheme Benefit (IISB) each year.^[4]
- Pleural plaque is not a prescribed disease.
- Even a tenuous history of asbestos exposure is accepted when a claim for mesothelioma is made.

People who believe they have had occupational exposure to asbestos often choose to make a civil claim for compensation against their former employer.

Treatment

Currently, treatment of asbestos related lung disease is largely symptomatic and supportive.

Associated diseases such as COPD and asthma are treated in the same as these conditions would be without history of asbestos exposure. **(See ‘COPD’ and ‘Asthma’ Protocols.)**

Medical Services

Oxygen therapy improves symptoms in the late stages of the diseases.

Treatments for mesothelioma include supportive care, surgery, radiotherapy, and chemotherapy but, individually, these have not improved survival. Recent trials suggest that combination treatment may offer an improvement in survival. Radical surgery followed by chemotherapy and radiotherapy has achieved better outcomes, especially for patients with local disease.^[8]

Prophylactic radiotherapy following invasive procedures (drainage or biopsy) has been found to reduce the risk of seeding along the track. A randomised study of 40 patients reduced the risk of seeding from 40% to zero. [13]

Pleural aspiration and pleurodesis help to relieve the breathlessness caused by pleural effusions.

New treatments such as photodynamic therapy, targeted cytokines and gene therapy are under investigation.^[8]

Prognostic scoring systems have been developed which are capable of stratifying patients into groups depending on their clinical characteristics and then predicting their likely survival. They help in the selection of participants for chemotherapy treatment trials.^[11]

The Primary Care Team and the Macmillan Service have a key role in providing palliative and supportive care for both the patient and their family. The MacMillan service is just one of many charities who, alongside the NHS, provide palliative care services for symptom control, psychosocial and spiritual support. Support is also available for carers and will continue following bereavement.

Prognosis

Asbestosis

Asbestosis is usually slowly progressive, with the speed of progression probably being related to the dose of asbestos to which the lungs have been subjected.^[4]

Asbestosis carries a five-fold increase in the risk of developing bronchial carcinoma.^[10]

There is evidence of a multiplicative increase in risk when asbestosis and smoking coexist.

40 – 50% per cent of smokers with asbestosis die of bronchial carcinoma.^{[4][5][10][11]}

Pleural Plaque

On its own, this condition carries a good prognosis.^[7]

Diffuse Pleural Thickening

This disease is usually slowly progressive.

Mesothelioma

Mesothelioma still has a very poor prognosis. Median survival is poor varying from 8-14 months in different studies. [14]

A few patients have indolent disease and may survive for up to 5 years.^[4]

Main Disabling Effects

The degree of disability caused by asbestos related lung disease depends on the particular condition. (From mild pleural plaque disease, which is asymptomatic, to mesothelioma, which is rapidly fatal.)

In non-malignant asbestos related lung disease, it is common to encounter concomitant conditions such as COPD and emphysema. These may add to the respiratory impairment.

Mesothelioma

Chest pain, breathlessness and weakness are the main disabling features of mesothelioma.

Non-malignant Asbestos Related Lung Disease

NB. Non-malignant asbestos related lung disease progresses slowly.

The primary disablement from non-malignant asbestos related lung disease is impaired exercise tolerance. Eventually, this might progress to the point that it limits 'walking quickly' and 'climbing flights of stairs.' In end stage disease, even 'washing' and 'dressing' may become difficult.

Assessing the Claimant

The assessment should be made using all the information available. This includes information from the claimant's file, informal observations, medical history, 'Typical Day', and examination.

- A key point to discover is whether the claimant is currently receiving active care from a Consultant.
If so, it suggests that the condition has been newly diagnosed, or that it has become worse.
If not, it suggests that the condition is stable, and that the rate of progression is slow.
This information shows the current severity of the condition, and helps to predict the prognosis.
- It is important to discover the claimant's reason for leaving employment.
The condition may have been diagnosed several years earlier, and it is likely that the claimant would have been able to continue at work. Was it due to respiratory impairment, an unrelated health problem, redundancy or some other circumstance?
- The focus of the assessment should be directed at the most disabling condition.

Medical Services

For example, in the case of a dock worker who received a diagnosis of pleural plaque 5 years ago, continued at work, but then suffered a back injury 8 months ago, the focus should be on the musculoskeletal system.

- A particular feature of asbestosis is **the breathlessness on sustained exertion**.

In the 'Typical Day' history, this might be illustrated by breathlessness on 'rushing for a bus', 'walking uphill', or 'climbing several flights of stairs.' Activities such as 'walking on level ground' or 'climbing a flight of twelve stairs' would be unlikely to cause significant problems.

- Variability is not a typical feature of non-malignant asbestos related lung disease.

If the claimant describes variability in their disability, then it may be due to concomitant disease or it may indicate inconsistency in their account.

In the IB-PCA, exemption is likely to have been granted to claimants with mesothelioma by the Decision Maker, or advised at the medical scrutiny stage. This condition is unlikely to be encountered during an IB-PCA examination session.

For those with non-malignant asbestos related lung disease, exemption should be considered if effort tolerance is severely limited, the claimant is using oxygen therapy, or they have had to adapt their home, for example by installing a stair lift or converting a room downstairs for their bedroom.

Psychological Effects

When workers are notified that due to occupational exposure to asbestos, they are at risk of severe chronic or malignant lung disease, psychological problems may result. Emotions such as anxiety or denial may affect their ability to process, remember, or act upon the information presented. Efforts to consider the psychological impact of notification have been made.^[12]

The knowledge that the diagnosis of asbestosis or diffuse pleural thickening may progress to respiratory failure or develop into a cancer can be difficult to come to terms with, and may lead to reactive depression. Similarly, the diagnosis of mesothelioma with its very poor prognosis can cause adjustment reactions not only for the patient, but also for their family. Adjustment reactions occur in about 30% of cancer patients.^[4]

Causal attribution, anger and the burden of legal proceedings may contribute to the suffering experienced by people with mesothelioma.^[15]

Reference List

1. Peto J, Hodgson JT, Matthews FE, Jones JR. Continuing increase in mesothelioma mortality in Britain. *Lancet* 1995;345:535-9.
2. Becklake MR. Symptoms and pulmonary functions as measures of morbidity. *Annals of Occupational Hygiene* 418;38:569-80.
3. Beritic-Stahuljak D, Valic F, Zuskin E. Relationship between cumulative occupational exposure to asbestos fibres and respiratory symptoms. *Acta Medica Croatica* 1991;45:283-95.
4. The Oxford Textbook of Medicine on CD ROM. Oxford University Press, 1996.
5. British Thoracic Society. Burden of Lung Disease. A statistics report from the British Thoracic Society. 2001.
6. Meyer JD, Holt DL, Chen Y, Cherry NM, McDonald JC. SWORD '99: surveillance of work-related and occupational respiratory disease in the UK. *Occupational Medicine (Oxford)* 2001;51:204-8.
7. Peacock C, Copley SJ, Hansell DM. Asbestos-related benign pleural disease. *Clinical Radiology* 2000;55:422-32.
8. Jaklitsch MT, Grondin SC, Sugarbaker DJ. Treatment of malignant mesothelioma. *World Journal of Surgery* 2001;25:210-7.
9. Zejda JE. Occupational exposure to dusts containing asbestos and chronic airways disease. *International Journal of Occupational Medicine & Environmental Health* 1996;9:117-25.
10. ABC of Work Related Disorders. 2nd Impression. BMJ Publishing Group, 2001.
11. Billings CG, Howard P. Asbestos exposure, lung cancer and asbestosis. *Monaldi Archives for Chest Disease* 2000;55:151-6.
12. Meyerowitz BE. Assessing quality of life when planning and evaluating worker notification programs: two case examples. *American Journal of Industrial Medicine* 1993;23:221-7.
13. Boutin C, Rey F, Viallet JR. Prevention of malignant seeding after invasive diagnostic procedures in patients with pleural mesothelioma. A randomised trial of local radiotherapy. *Chest* 1995; 108:754-8
14. British Thoracic Society Standards of Care Committee. Statement on malignant mesothelioma in the United Kingdom (BTS statement). *Thorax* 2001; 56(4): 250-265
15. Clayson H. Suffering in mesothelioma: concepts and contexts. *Progress in Palliative Care* 2003;11 :251-254.

Bibliography

The Oxford Textbook of Medicine 3rd Edition. CD-ROM. Oxford University Press.
Asbestosis. Edited by A Seaton.
Mesothelioma. Edited by MK Benson.
Lecture Notes in Respiratory Medicine 5th Edition. SJ Bourke et al Blackwell Scientific.
ABC of Work Related Disorders. 2nd Impression 2001. Edited by David Snashall. BMJ Publishing Group.
Practical Pulmonary Rehabilitation. Edited by M Morgan, S Singh. Chapman and Hall Medical.
IARC monographs on the evaluation of carcinogenic risks to humans. 1972-1996 Vol. 1-66. International Agency for Research on Cancer. Lyons.
Burden of Lung Disease. A statistics report from the British Thoracic Society. Nov. 2001.