

## **Cryptogenic Organising Pneumonia**

*Dr Nicole Goh Consultant Respiratory Physician Austin Hospital, Victoria*

**Description:** Pneumonia is an inflammation (collection of immune cells) of the lungs. The alveoli (or air sacs) are typically inflamed and are abnormally filled with fluid. "Organizing" refers to unresolved pneumonia in which excessive healing tissue is laid down. Therefore, the term "organizing pneumonia" (OP) [previously known as bronchiolitis obliterans organizing pneumonia (BOOP)] is a condition caused by inflammation and excessive proliferation of healing tissue within the alveoli, and also the bronchioles (small airways). OP can be seen in association with a number of conditions including connective tissue diseases (e.g. rheumatoid arthritis, polymyositis and dermatomyositis), certain medications, bone marrow transplantation and radiation therapy. The term "cryptogenic organizing pneumonia" (COP) is used when no cause can be found.

**Epidemiology and diagnosis:** Most patients with OP will have symptoms for less than two months before seeking medical attention. Symptoms include a flu-like illness, cough, fever, a feeling of illness (malaise), fatigue, shortness of breath, and occasionally, weight loss. There are no specific abnormalities on routine laboratory tests or on physical examination, although crackling sounds in the chest can sometimes be heard with a stethoscope. Patients are often treated as having infectious pneumonia, and the diagnosis is only suspected when no infectious agents are found, and there has been no response to multiple antibiotics.

Initial investigations include a chest x-ray, lung function tests and blood tests. Chest x-ray typically shows white patches in the lungs in more than one area. The term "consolidation" is given to these changes. Lung function tests usually show that the amount of air the lungs can hold is below normal. The amount of oxygen in the blood can be low at rest and even lower with exercise. Although there are no specific abnormalities on blood tests, markers indicating the presence of inflammation are often high.

Occasionally, markers of a connective tissue disease might also be present on blood tests. A CT (computed tomography) scan of the chest is often required and this provides more detail compared to the chest x-ray. A bronchoscopy (a telescope into the airways) may be required to obtain samples to exclude infection or to obtain a small sample of the lung tissue. Very occasionally, a larger sample of the lung tissue may be required and patients will need to undergo surgery for this.

**Management:** OP may resolve spontaneously; however, in most cases, patients will require treatment to decrease the amount of inflammation. Most patients recover with treatment, with

symptoms improving within days or weeks. Radiographic findings demonstrate improvement in the majority of patients; however, the disease may persist in a minority of patients despite treatment. In about 30% of patients (usually those with a CTD), the disease comes back when treatment is stopped. Patients with OP generally respond well to treatment with steroids. In some patients, immunosuppressants (drugs that dampen the immune system) may also be required.

Rarely, organising pneumonia can progress rapidly to respiratory failure with requirement for ventilation and death has been reported in this setting. This is most likely to occur in patients with polymyositis or one of the anti-synthetase syndromes. Even if treatment arrests the inflammatory phase of the illness, some lung scarring usually results and patients may be left with reduced exercise tolerance and permanent changes on CXR. Long term follow-up is recommended in view of the possibility of relapse.

### **References:**

*Cordier JF. Cryptogenic organizing pneumonia. Good review article of COP with particular focus on aetiology and pathogenesis. Eur Respir J 2006 Aug;28(2):422-46.*

*Davison AG, Heard BE, McAllister WA, Turner-Warwick ME. Cryptogenic organizing pneumonitis. First paper to describe the clinicopathological syndrome of COP. Q J Med 1983;52(207):382-94.*

*Oikonomou A, Hansell DM. Organizing pneumonia: the many morphological faces. Describes some of the less characteristic radiological appearances of COP. Eur Radiol 2002;12(6):1486-96.*

### **Useful Links**

For looking up drugs that may cause OP: [www.pneumotox.com](http://www.pneumotox.com)







This brochure is one in a series produced by Lung Foundation Australia to provide information on lung disease, its treatment and related issues. The information published by Lung Foundation Australia is designed to be used as a guide only, is not intended or implied to be a substitute for professional medical treatment and is presented for the sole purpose of disseminating information to reduce lung disease.

Any information relating to medication brand names is correct at the time of printing. Lung Foundation Australia has no control or responsibility for the availability of medications, which may occasionally be discontinued or withdrawn.

Please consult your family doctor or specialist respiratory physician if you have further questions relating to the information contained in this leaflet. For details of patient support groups in Australia please call 1800 654 301.