BMJ Best Practice

Organising pneumonia

Straight to the point of care

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Summary

Organising pneumonia (OP) is an inflammatory disorder involving both the peripheral bronchioles and alveoli simultaneously. It has distinctive radiographic findings, histological features, and response to corticosteroids (unlike usual interstitial pneumonia).

OP may be caused by multiple insults such as medication, infection, rheumatological disease, autoimmune disease, post-transplantation, radiation, and environmental causes. In cryptogenic organising pneumonia, a cause cannot be elicited after a careful history, examination and pertinent laboratory studies.

High-resolution chest computed tomography scan shows bilateral patchy triangular ground glass opacities with air bronchograms usually located peripherally.

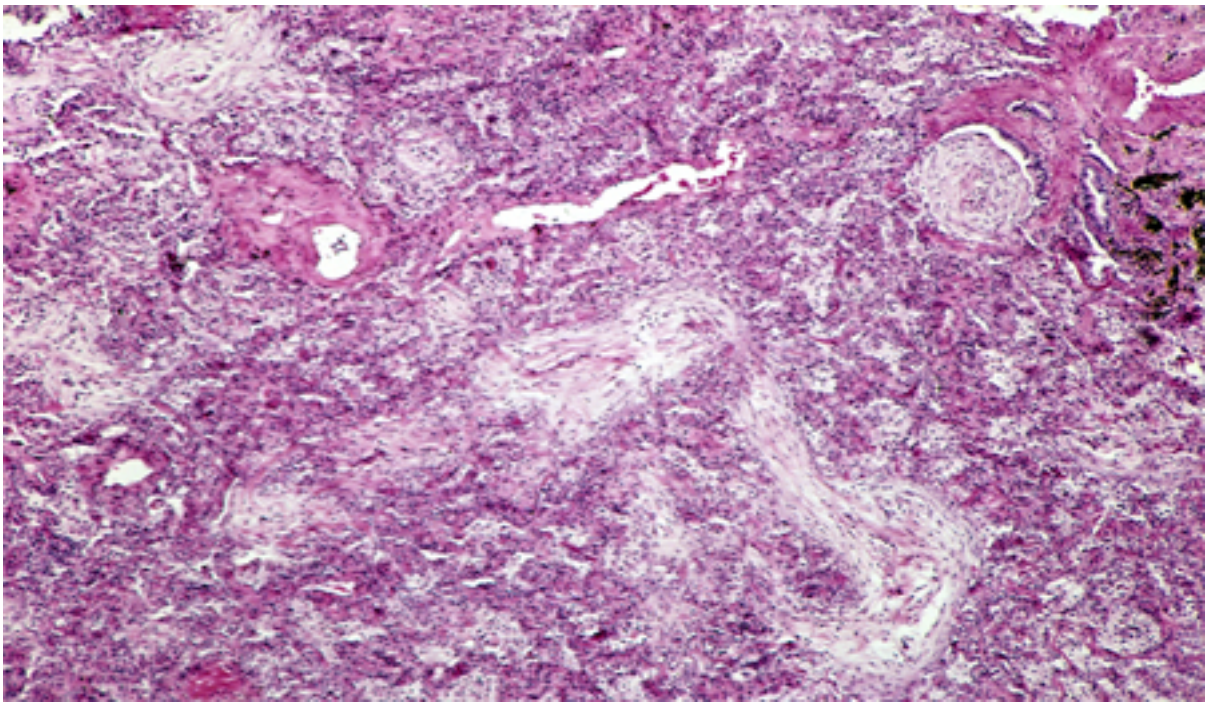
Most often, diagnosis is made using clinico-radiological criteria and usually in the setting of a multidisciplinary team. However, lung biopsy may be required to establish the definitive diagnosis in patients with unusual findings.

Cryptogenic OP may require treatment, and prednisolone is the most commonly used drug.

For patients with secondary OP, primary management is removal of the inciting cause or treatment of the underlying disease. In some patients, corticosteroids may be indicated.

Definition

Organising pneumonia (OP) is defined as organised polypoid granulation tissue in the terminal bronchioles, alveolar ducts and alveoli; it has distinctive radiographic findings, histological features, and response to therapy.[1] [2] [3] The term cryptogenic is applied when a causative factor cannot be found.



Medium-powered pathology slide showing circular and branching bronchioles filled with polypoid plugs of granulation tissue and alveoli filled with organising pneumonia

From the collection of Gary R. Epler, MD

OP was previously called bronchiolitis obliterans with organising pneumonia (BOOP) but this led to some confusion with the entirely separate disease bronchiolitis obliterans.[4] Hence, a revision of the nomenclature to organising pneumonia, cryptogenic and secondary. Some clinicians still adhere to the earlier nomenclature. The term cryptogenic organising pneumonia (COP) is a general term referring to organised inflammatory process in the alveoli from an unknown cause.[5] COP is the preferred term because it captures an 'acinar' rather than an airway disease, and BOOP may be confused with obliterative bronchiolitis.[4]

The following are a few differences between obliterative bronchiolitis, where there is obstruction of the bronchioles due to inflammation, and organising pneumonia. Wheezing is not a common symptom of OP; crackles, and not wheezes, are heard by auscultation in OP. The FEV1/FVC ratio is normal or slightly increased in OP, not decreased as in airway obstructive diseases and the radiographic findings show bilateral patchy infiltrates, not normal or hyperinflation seen in airflow obstructive diseases.

Epidemiology

The incidence of organising pneumonia (OP) is somewhat uncertain and estimates are commonly drawn from registries which may result in an overestimate of disease incidence but is between 0.5 to 1.5 per 100,000.[31] [32] The incidence of OP is estimated at 1.97 per 100,000 in Iceland and in the rest of the world ranges from 0.2 to 1.45 per 100,000 or is thought to be approximately 5% of interstitial lung disease patients noted in these registries.[31] The fact that the data is sourced from registries may result in some inaccuracy. The incidence does not differ significantly worldwide.[33] OP occurs equally among men and women, and is not related to smoking. In the Iceland dataset, cryptogenic OP had an incidence of 1.1 per 100,000 while secondary OP had an incidence of 0.87 per 100,000.[31] A similar trend is reflected in an Italian study.[34] In other studies, however, the incidence of secondary OP was as high as 87% of the total number of patients presenting with OP.[35] It is noteworthy that in a Chinese study, all patients had follow up by phone calls for 4 years after inclusion in study, and 4% of those originally classified as cryptogenic OP had reclassification as secondary OP.[36]

Aetiology

Organising pneumonia (OP) can be cryptogenic, where no cause can be found, or secondary to a known factor. Possible causes of secondary OP are listed below.

Infections

- *Chlamydia* , *Legionella* , and *Mycoplasma*
- Adenoviridae, *Cytomegalovirus* , and influenza virus
- Malaria
- *Pneumocystis* [10]
- *Cryptococcus* .

Medications[11]

- Antibiotics: amphotericin-B, cephalosporins, minocycline, nitrofurantoin[12]
- Cardiovascular drugs: amiodarone, acebutolol
- Cancer chemotherapy drugs: bleomycin, busulfan, methotrexate, doxorubicin, thalidomide, cytosine-arabinoside (ARA-C), cytarabine, chlorambucil, rituximab
- Anti-inflammatory agents: gold, sulfasalazine, mesalazine, bucillamine, infliximab
- Immunosuppressive agents: azathioprine, mercaptopurine, tacrolimus, sirolimus, everolimus
- Anticonvulsants: carbamazepine, phenytoin
- Miscellaneous drugs: interferon, ticlopidine, L-tryptophan, risedronate, illicit use of cocaine.

Rheumatological or connective tissue disorders

- Rheumatoid arthritis[13] [14]
- Lupus erythematosus
- Sjogren's syndrome
- Sweet's syndrome[15]
- Polymyositis/dermatomyositis
- Scleroderma-progressive systemic sclerosis
- Ankylosing spondylitis
- Polymyalgia rheumatica
- Behcet's disease.

Immunological disorders

- Common variable immunodeficiency syndrome
- Essential mixed cryoglobulinaemia.

Organ transplantation

- Lung[16]
- Bone marrow[17] [18]
- Kidney and liver.

Radiotherapy

- Post breast radiation (1% to 3% of patients).[19] [20] [21] [22]

Environmental or occupational exposures

- Textile printing dye
- *Penicillium* mould dust
- House fire
- Food spice processing[23]
- Paraffin mineral oil.[24]
- Vaping.[25]

Miscellaneous

- Inflammatory bowel disease
- HIV infection
- Illicit use of cocaine
- Myelodysplastic syndrome
- Hunner's interstitial cystitis
- Chronic thyroiditis and alcoholic cirrhosis
- Seasonal syndrome with cholestasis
- Primary biliary cirrhosis
- Coronary artery bypass graft surgery
- Cancer and haematological malignancy[26] [27]
- Aspiration[28] [29]
- Hydatid cyst.[30]

Pathophysiology

Organising pneumonia (OP) is an inflammatory lung disease caused by a specific cascade of cytokine events. It differs from the inflammation occurring in asthma and chronic bronchitis. The pathogenesis is not a fibrotic process as in usual interstitial pneumonia (UIP). Naturally timed apoptosis may be an important distinction between OP (an inflammatory process) and UIP (a fibrotic process), because apoptotic activity is increased in the fibromyxoid connective tissue of OP but not UIP.[37] The cytokine profile of OP shows an increased degree of macrophage and lymphocyte activation with the T-1 response.[38] In the reovirus model, T cells have an important role in the pathogenesis of OP, demonstrated by the fact that depletion of CD4+ or CD8+ cells and treatment with corticosteroids decrease expression of the proinflammatory and profibrotic cytokines.[39]

On low to medium power light microscopy, the epithelial buds, consisting of granulation tissue obtruding into the distal airspaces, can be seen. Fibrin and proliferating fibroblasts and myofibroblasts comprise the cellular matrix. Cryptogenic OP presents in three main patterns:

- Multiple patchy alveolar opacities in a perilobular distribution
- Focal nodules
- Diffuse infiltrative opacities that are peripheral and bilateral.

Less common patterns are the reverse halo (atoll) sign which consists of an area of inflammation with central clearing, bandlike opacities or crazy paving.[32] [40] Over time, a fibrotic pattern may appear with subpleural reticulations and mild volume loss.

Classification

Clinical classification[3]

OP can be classified according to clinico-radiological pattern:

- Idiopathic OP, i.e,. cryptogenic organizing pneumonia (COP) (no underlying cause established)[1] [2] [3]
- Rapidly progressive OP[6] [7]
- Focal nodular OP[8] [9]
- Secondary OP

Secondary OP can be classified according to aetiology:

- Post-infectious OP
    - *Chlamydia* , *Legionella* , *Mycoplasma*
    - Adenoviridae, *Cytomegalovirus* , and influenza virus
    - Malaria
    - *Pneumocystis* [10]
    - *Cryptococcus*
    - COVID-19 (severe acute respiratory syndrome coronavirus 2 [SARS CoV-2]).
- Drug-related OP[11]
    - Antibiotics: amphotericin-B, cephalosporins, minocycline, nitrofurantoin[12]
    - Cardiovascular drugs: amiodarone, acebutolol
    - Cancer chemotherapy: bleomycin, busulfan, methotrexate, doxorubicin, thalidomide, cytosine-arabinoside (ARA-C), cytarabine, chlorambucil, rituximab
    - Anti-inflammatory agents: gold, sulfasalazine, mesalazine, bucillamine, infliximab
    - Immunosuppressive agents: azathioprine, mercaptopurine, tacrolimus, sirolimus, everolimus
    - Anticonvulsants: carbamazepine, phenytoin
    - Miscellaneous drugs: interferon, ticlopidine, L-tryptophan, risedronate, cocaine (illicit use).
- Rheumatological or connective tissue OP
    - Rheumatoid arthritis[13] [14]
    - Lupus erythematosus
    - Sjogren's syndrome
    - Sweet's syndrome[15]
    - Polymyositis, dermatomyositis

    - Scleroderma-progressive systemic sclerosis
    - Ankylosing spondylitis
    - Polymyalgia rheumatica
    - Behcet's disease.
- Immunological disorder OP
    - Common variable immunodeficiency syndrome
    - Essential mixed cryoglobulinemia.
- Organ transplantation OP
    - Lung[16]
    - Bone marrow[17] [18]
    - Kidney and liver.
- Radiotherapy OP
    - Following breast radiation (1% to 3% of patients).[19] [20] [21] [22]
- Environmental or occupational exposures
    - Textile printing dye
    - *Penicillium* mould dust
    - House fire
    - Food spice processing[23]
    - Paraffin mineral oil.[24]
    - Vaping.[25]
- Miscellaneous OP
    - Inflammatory bowel disease
    - HIV infection
    - Illicit use of cocaine
    - Myelodysplastic syndrome
    - Hunner's interstitial cystitis
    - Chronic thyroiditis and alcoholic cirrhosis
    - Seasonal syndrome with cholestasis
    - Primary biliary cirrhosis
    - Coronary artery bypass graft surgery
    - Cancer and haematological malignancy[26] [27]
    - Aspiration[28] [29]
    - Hydatid cyst.[30]

Case history

Case history #1

A 48-year-old school teacher develops a flu-like illness with low-grade fever, mild cough, and generalised malaise. Physical examination shows bilateral end-inspiratory crackles. The chest x-ray shows bilateral patchy infiltrates. A 10-day course of antibiotics does not improve the symptoms, and the antibiotic is changed to a fluoroquinolone. Shortness of breath develops, and the high-resolution chest computed tomography (CT) scan shows bilateral ground glass opacities with air bronchograms, some triangular in

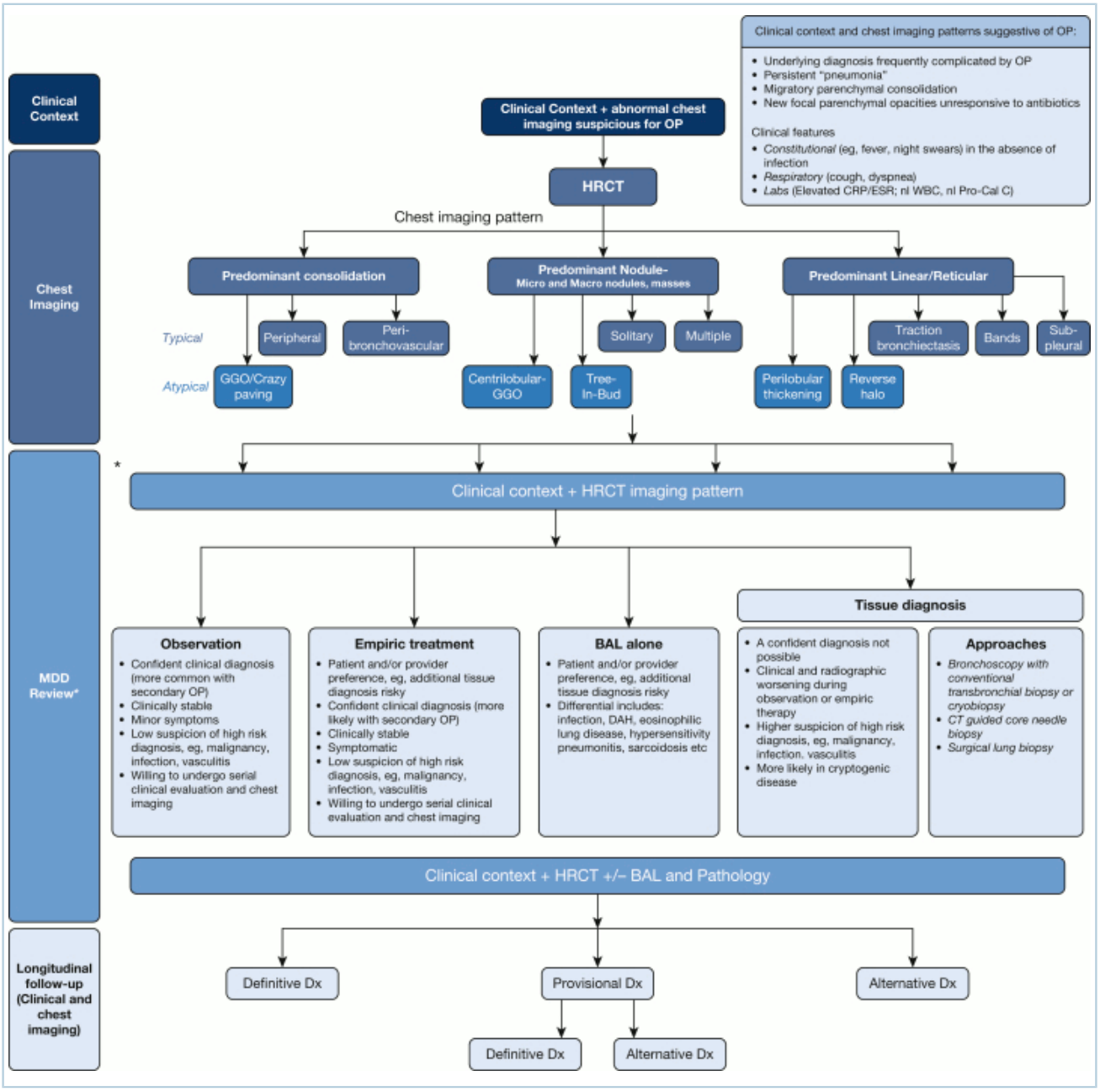
shape. The vital capacity is decreased to 72% predicted, the FEV1/FVC ratio is normal at 81%, and the diffusing capacity is decreased to 58% predicted.

Case history #2

A 60-year-old man with a 70 pack year history of smoking goes for low dose chest CT as part of his lung cancer screening. Chest CT shows a 4 cm x 3 cm right lower lobe mass in a subpleural location. On history, the patient reports having a 'walking pneumonia' 4 months earlier which was treated by his family doctor with doxycycline for 7 days and albuterol inhaler for wheezing. He denies cough, fever, sputum or weight loss. He is back to his baseline exercise tolerance, walking half a mile with mild wheezing at the end but rapid recovery not requiring inhaler therapy. He had an initial chest x-ray at the diagnosis of pneumonia but felt well after antibiotics and has not followed with his family doctor. PET scan showed uptake in the right lower lobe mass only with a standard uptake value (SUV) of 3.1. The patient is referred for a transthoracic biopsy.

Approach

The presentation of OP depends on the type. To establish whether there is any underlying cause, it is essential to take a full medical, surgical, and drug history and consider any possible occupational or environmental exposures. Most often, diagnosis is made using clinico-radiological criteria and usually in the setting of a multidisciplinary team.



Algorithmic approach to organizing pneumonia. # A formal MDD may not be required in all cases, especially if the combination of clinical context and radiographic pattern is sufficiently convincing of the OP diagnosis. In such cases, a discussion between the physician and the radiologist is strongly encouraged. CRP = C-reactive protein; DAH = diffuse alveolar hemorrhage; Dx = diagnosis; ESR = erythrocyte sedimentation rate; GGO = ground-glass opacification; HRCT = high-resolution CT; MDD = multidisciplinary discussion; nl Pro-Cal C = normal procalcitonin; nl WBC = normal WBC; OP = organizing pneumonia

Cherian SV, et al. Chest. 2022 Jul;162(1):156-78. doi: 10.1016/j.chest.2021.12.659. Epub 2022 Jan 14; used with permission

Clinical diagnosis

Cryptogenic OP

- Typically presents as a flu-like illness with mild fever, arthralgia, fatigue lasting for several days, and a mild cough lasting 1 to 3 months

- Shortness of breath develops later as increasing numbers of alveoli are affected
- Bilateral end-inspiratory crackles can be heard
- There may be a rapidly progressive subtype presenting as bilateral infiltrates and acute hypoxic respiratory failure often requiring mechanical ventilation.

Focal nodular OP

- May be seen on chest x-ray or chest computed tomography (CT) scan as an incidental finding during investigation of an unrelated cough or chest pain
- Nodule may be circular in 1 lung, or there may be 3 to 5 nodules in both lungs
- May cause mild respiratory symptoms but is often asymptomatic
- Typically low positive uptake on positron emission tomography (PET) scan[42]
- Diagnosis often established after resection of a pulmonary mass thought to be a neoplastic process preoperatively.

Post-infectious OP

- Develops in people who have an infectious pneumonia that initially responds to antibiotics, but the patchy infiltrates and mild respiratory symptoms persist as the inflammatory component of the infection becomes organised. Experience from the coronavirus disease 2019 (COVID-19) pandemic supports this observation.[43] [44]

Drug-related OP

- Occurs several weeks after medication is started
- Presents with bilateral patchy infiltrates and cough that mimic infection or sometimes neoplasm, both of which must be ruled out before beginning corticosteroid therapy
- Biopsy may be needed.

Connective tissue-related OP

- Generally occurs months to years after diagnosis of the underlying disorder, although OP with cough, shortness of breath, and bilateral patchy infiltrates may precede the connective tissue disorder by months or even 1 to 2 years in people with polymyositis.[45] [46]

OP following lung transplantation

- Acute fibrinous organizing pneumonia is a feared postoperative complication of lung transplantation. This is associated with poor survival. Survivors may develop bronchiolitis obliterans syndrome.[47]

OP following breast radiation

- Has emerged as an important development
- May occur in up to 2% to 3% of women
- Risk factors include age ≥50 years and concurrent endocrine therapy in women[21]
- Tamoxifen not a risk factor
- Mean latency is 4.4 months after radiation with a range of 2.3 to 7.9 months.

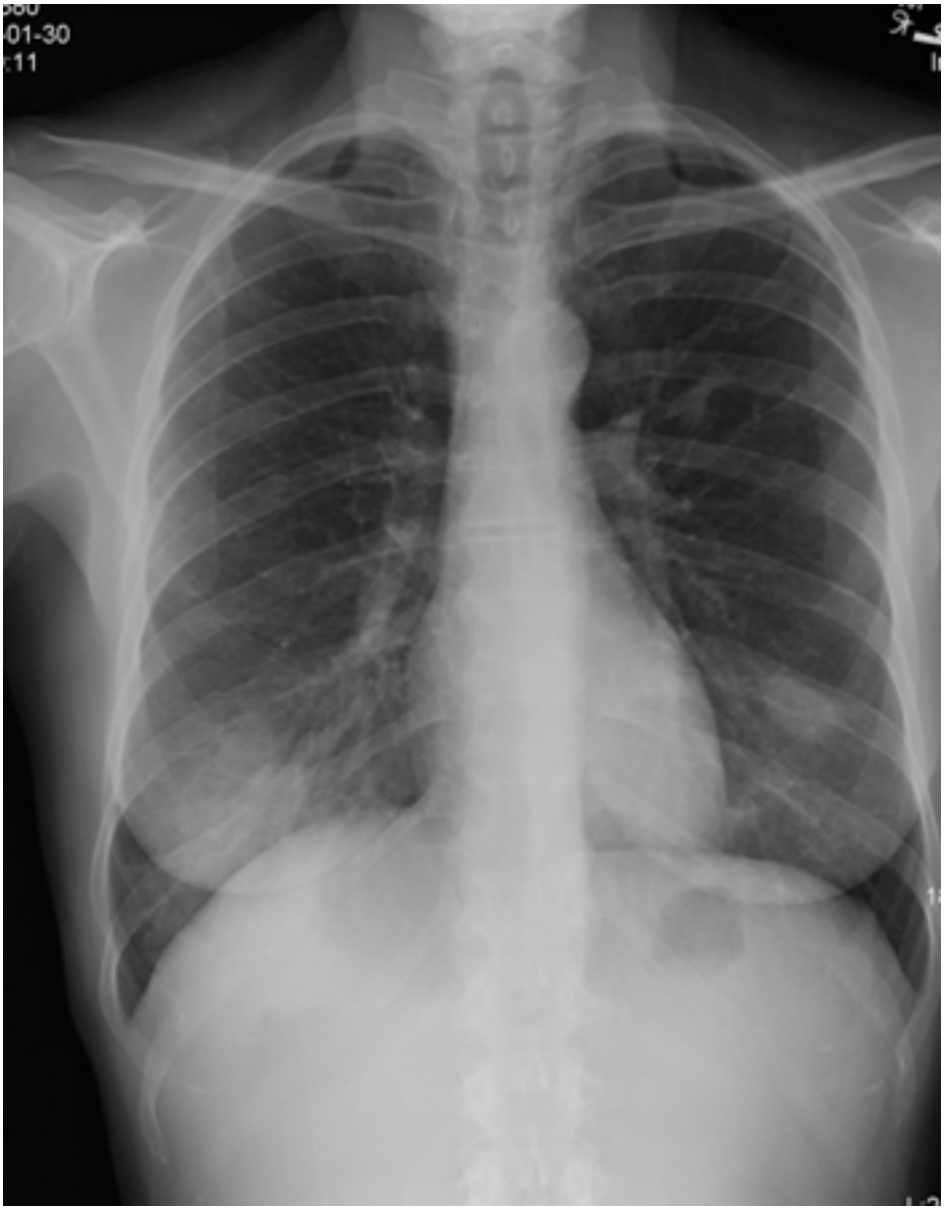
OP following occupational or environmental exposure

- Occupational and environmental history important to exclude potential exposures
- Occupational information includes job title and exposure information
- Environmental information includes unusual toxic home exposures
- Toxins that may trigger OP include textile printing dye, *Penicillium* mould dust, house fire, food spice processing, and paraffin mineral oil.[23] [24] [41]

Investigations

Chest x-ray is the initial investigation of choice and usually shows patchy infiltrates.[48] [49] In many cases, patients are misdiagnosed with a community acquired pneumonia and are treated with a course of antibiotics.

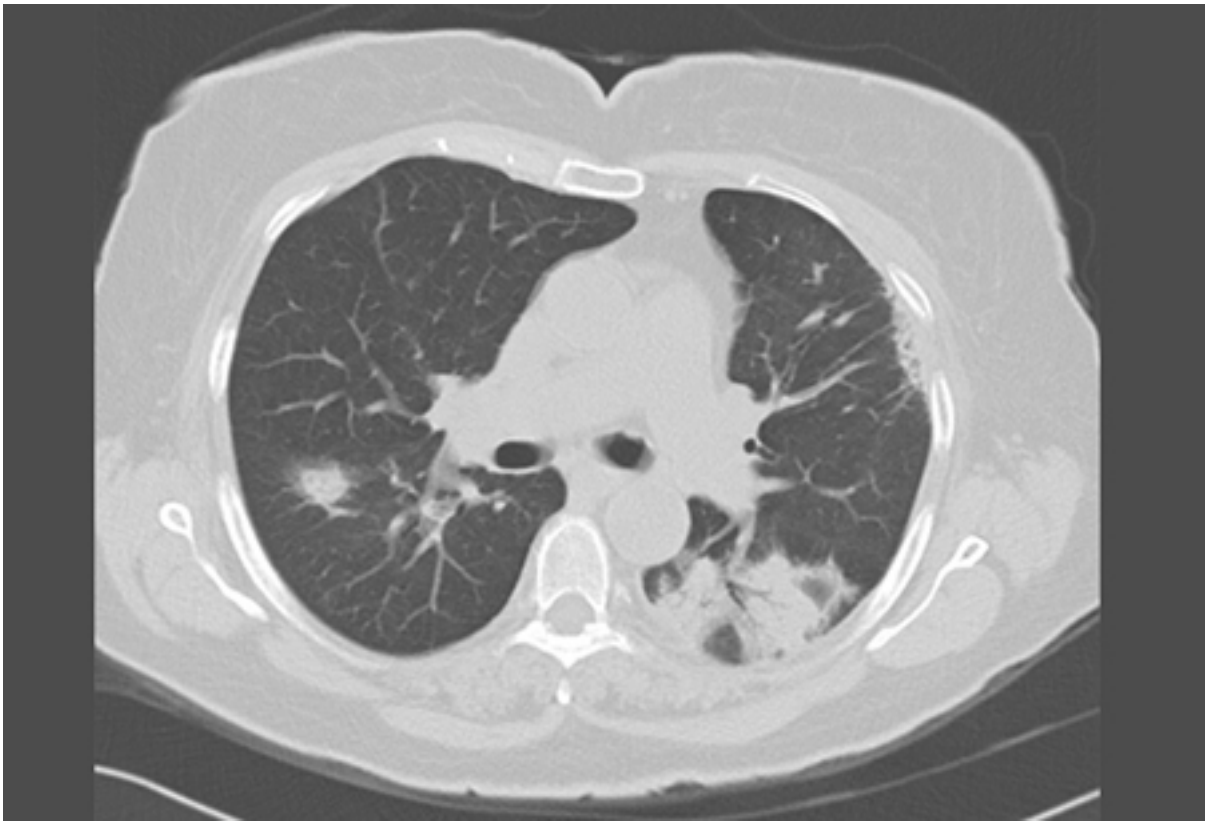
Quite often, the symptoms persist and the chest x-ray worsens, usually with enlargement of the patchy infiltrates. In some situations, waxing and waning opacities may develop over a couple of weeks.



Chest x-ray showing bilateral patchy infiltrates

From the collection of Gary R. Epler, MD

A high-resolution chest computed tomography (CT) scan often shows bilateral ground glass opacities with air bronchograms.[5] [48][49] In OP, the ground glass opacities with the air bronchograms are often in the shape of a triangle with the base of the triangle along the pleural surface and the apex towards the mediastinum ('the triangle sign').[2] Other radiographic patterns such as a diffuse infiltrative pattern, atoll sign, crazy paving, fibrotic changes and bands may be seen.



High-resolution chest CT showing bilateral ground glass opacities and a posterior triangular-based infiltrate with an air bronchogram

From the collection of Gary R. Epler, MD

Pulmonary function tests (PFTs), if obtained, will show a decreased vital capacity and diffusing capacity with no airflow obstruction.[2] [3] [6]

Blood studies may show an increased WBC and erythrocyte sedimentation rate. C-reactive protein measurement, sputum culture and atypical viral screen may all help to exclude infection.[50]

A general autoimmune and myositis profile should be performed to screen for connective tissue disease if there is no obvious cause for OP as it may precede the diagnosis of connective tissue diseases by several months or years. The profile should include testing for: antinuclear antibodies, rheumatoid factor, anti-Scl-70 antibodies, anti-Sjögren-syndrome-related antigen A antibodies, anti-Sjögren-syndrome-related antigen B antibodies, cyclic citrullinated peptide antibodies, anti-ribonucleoprotein antibodies, antisynthetase antibodies, anticardiolipin antibodies, lupus anticoagulant antibodies, anti-glomerular basement membrane antibodies, and antineutrophil cytoplasmic antibodies/myeloperoxidase antibodies/proteinase 3 antibodies. Creatine kinase is useful for screening for polymyositis OP.

Often, a diagnosis of OP may be made based on clinical history, typical radiographic findings and significant negative or positive results from appropriate blood studies. Bronchoscopic investigations, including analysis of bronchoalveolar lavage fluid, may help to rule out other differential diagnoses such as eosinophilic pneumonia and to confirm that there is no active infection. The value of transbronchial biopsy is often doubtful since there is often insufficient tissue for the architecture to be evaluated, or the presence of crush artifacts which diminish its value. In one single center study, only 7 of 34 transbronchial biopsies demonstrated the pathology.[51]

Some experts feel that pathological diagnosis is necessary. In that respect, a surgical lung biopsy represents best practice for confirmation in an atypical situation such as rapidly progressive disease, diffuse infiltrative disease, no known aetiology or associated disorder, an unusual chest CT pattern, or with moderately severe symptoms. Pathological criteria also state that the following features should not be present: extensive interstitial fibrosis, traction bronchiectasis, and histological honeycombing.[1] [2] Video-assisted thoracoscopy (VAT) is the preferred method for obtaining tissue.[28]

CT-guided transthoracic needle biopsy and transbronchial lung cryobiopsy are minimally invasive procedures that can be used as alternatives to surgical lung biopsy.[52] [53] [54] These may be important options in elderly or frail patients, as well as those with focal lesions. However, both procedures produce relatively small biopsy specimens that must be interpreted in the context of associated clinical and radiological findings.[53]

History and exam

Key diagnostic factors

presence of risk factors (common)

- Risk factors include infectious pneumonia, use of certain medications, history of connective tissue or immunological diseases, recent organ transplantation, breast radiotherapy, and exposure to environmental toxins.

flu-like illness with low-grade fever, fatigue, and arthralgia (common)

- Common with cryptogenic OP.
- Symptoms last for several days, typically <2 months.

Other diagnostic factors

cough (common)

- Cough is usually mild with no sputum production.

shortness of breath (common)

- Not a feature early in the disease but develops as OP becomes involved with the lung parenchyma.

bilateral crackles (common)

- End-inspiratory bilateral crackles occur in up to 80% of patients with OP.
- Wheezes do not occur.

Risk factors

Strong

infectious pneumonia

- OP may occur after infectious pneumonias. Organisms include viral agents, bacterial agents, atypical organisms, and parasites.[10]

connective tissue diseases

- OP may occur in association with all of the connective tissue disorders, including rheumatoid arthritis, lupus erythematosus, polymyositis and dermatomyositis, Sjogren's syndrome, mixed connective tissue, and antiphospholipid syndrome.[13] [14] In some situations, the drug used for treating the connective tissue disease can also cause OP (e.g., gold or methotrexate for treatment of rheumatoid arthritis).

immunological diseases and inflammatory bowel disease

- OP occurs in association with immunological diseases such as common variable immunodeficiency syndrome and inflammatory bowel disease. Sulfasalazine used for treatment of inflammatory bowel disease may also cause OP.

organ transplantation

- OP can occur in up to 10% of lung and bone marrow transplant recipients.[16] [17] OP has been reported in liver and kidney recipients.

medication use

- Over 30 medications can cause OP, including antibiotics, cardiovascular drugs (e.g., amiodarone), cancer chemotherapy drugs (e.g., bleomycin), anti-inflammatory agents, immunosuppressive agents, and anticonvulsants.

breast radiotherapy

- OP following breast radiotherapy may occur in 1% to 3% of patients and may resolve without corticosteroid treatment.[19] [20] [21] [22]

exposure to toxins

- Documented reports of OP caused by occupational or environmental toxic exposure are rare. Textile printing dye has been the most well documented, and others include *Penicillium* mould dust, house fire, food spice processing, and paraffin mineral oil.[23] [24] [41]

Weak

vaping

- Many of the flavouring agents in e-liquids have been associated with sporadic case reports of lung injury, including OP, due to e-cigarette or vaping product use.[25] The pattern of acute lung injury is typical of OP, diffuse alveolar damage, or both.[25]

Investigations

1st test to order

Test	Result
chest#x-ray • The most important initial diagnostic test for OP.[48] [49] • Chest x-ray is also of use in evaluating alternative diagnoses.[48] [49]	**bilateral patchy infiltrates; waxing and waning opacities may develop over a couple of weeks; focal OP may present as a circular nodule in 1 lung or 3 to 5 nodules in both lungs**
high-resolution chest CT scan (HRCT) • HRCT is fundamental to establishing a diagnosis of OP and can be used to monitor the patient for treatment or response to treatment.[48] [49] • The 'triangle sign' (the triangular ground glass opacity with the base on the pleura and the apex towards the mediastinum) is often distinctive for OP.[2]	**bilateral patchy ground glass opacities with air bronchograms usually located peripherally; focal OP may present as circular nodule in 1 lung or 3 to 5 nodules in both lungs; other radiographical patterns such as a diffuse infiltrative pattern, atoll sign, crazy paving, fibrotic changes and bands may be seen in OP**
FBC • Non-specific finding.	**WBC may be normal but often increased to 10,000-15,000/microlitre in cryptogenic OP**
erythrocyte sedimentation rate • Non-specific finding.	**elevated**

Other tests to consider

Test	Result
CRP • May help to exclude infection.	**raised in infection**
sputum culture • May help to exclude infection.	**may identify pathogen associated with infection**
atypical viral screen • May help to exclude infection.	**may identify virus associated with infection**
pulmonary function tests • In addition to the vital capacity and lungs volumes, obtaining the diffusing capacity is important to determine severity and monitor disease course.	**decreased vital capacity and diffusing capacity with no airflow obstruction**
surgical lung biopsy • Should be strongly considered especially in patients with an atypical presentation such as rapidly progressive disease, diffuse infiltrative disease, no known aetiology or associated disorder, an unusual chest CT pattern, or with moderately severe symptoms. Pathological criteria also state that the following features should not be present: extensive interstitial fibrosis, traction bronchiectasis, and histological honeycombing.[1] [2] • A video-assisted thoracoscopy procedure is the preferred method for obtaining tissue.[28] • CT-guided transthoracic needle biopsy and transbronchial lung cryobiopsy are minimally invasive procedures that can be used as alternatives to surgical lung biopsy.[52] [53] [54] These may be important options in elderly or frail patients, as well as those with focal lesions. However, both procedures produce relatively small biopsy specimens that must be interpreted in the context of associated clinical and radiological findings.[53]	**organised polypoid granulation inflammatory tissue in the distal bronchial airways, respiratory bronchioles, alveolar ducts, and alveoli**
creatine kinase • This test should be obtained if there is no obvious cause, because OP may precede the diagnosis of connective tissue diseases by several months or years. Creatine kinase is useful for screening for polymyositis OP.	**usually negative; elevated if polymyositis is underlying cause**
general autoimmune and myositis profile • Should be performed to screen for connective tissue disease if there is no obvious cause for OP as it may precede the diagnosis of connective tissue diseases by several months or years. The profile should include testing for: antinuclear antibodies, rheumatoid factor, anti-Scl-70 antibodies, anti-Sjögren-syndrome-related antigen A antibodies, anti-Sjögren-syndrome-related antigen B antibodies, cyclic citrullinated peptide antibodies, anti-ribonucleoprotein antibodies, antisynthetase antibodies, anticardiolipin antibodies, lupus anticoagulant antibodies, anti-glomerular basement membrane antibodies, and antineutrophil cytoplasmic antibodies/ myeloperoxidase antibodies/proteinase 3 antibodies.	**elevated antibodies if connective tissue disease is underlying cause**
positron emission tomography (PET) scan • May be useful in some patients thought to have focal nodular OP.	**typically low positive uptake on PET scan**

Test	Result
bronchoscopy • Bronchoscopic investigations, including analysis of bronchoalveolar lavage fluid, may be useful in some patients thought to have OP.	**may help to rule out other differential diagnoses such as eosinophilic pneumonia and to confirm that there is no active infection**

Differentials

Condition	Differentiating signs / symptoms	Differentiating tests
Nodular sarcoidosis	• No particular differentiating signs and symptoms.	• CXR may show enlarged hilar lymph nodes.
Chronic eosinophilic pneumonia (CEP)	• No particular differentiating signs and symptoms.	• High-resolution chest CT scan (HRCT) in CEP shows peripheral ground glass opacities with sparing of the central portions of the lungs. • FBC with differential may show increased blood eosinophils. • Bronchoalveolar lavage with eosinophil count in excess of 30%.
Non-specific interstitial pneumonia (NSIP)	• Shortness of breath will be progressive and crackles will persist while the patient is receiving corticosteroid therapy.	• HRCT in NSIP shows ground glass opacities +/- traction bronchiectasis and honeycombing with subpleural sparing.[55]
Idiopathic pulmonary fibrosis with a usual interstitial pneumonia (UIP) pattern	• Patients with UIP will have progressive shortness of breath and increasing degree of bilateral crackles.	• HRCT shows linear opacities at the lung bases, traction bronchiectasis, and subpleural honeycombing.
Granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis)	• Patients with granulomatosis with polyangiitis (formerly known as Wegener's granulomatosis) may have nasal-sinus and renal symptoms.	• HRCT shows bilateral multiple cavitary lesions. • A positive ANCA is typically present.
Acute interstitial pneumonia	• Symptoms and findings same as rapidly progressive OP.	• HRCT may show disrupted lung architecture with traction bronchiectasis and early honeycombing.
Pulmonary metastasis and primary adenocarcinoma	• Symptoms of cough and shortness of breath progress when the patient is treated with corticosteroids.	• Kerley B lines occur in carcinomatosis. • Chest x-ray may detect a solitary pulmonary nodule, mass, pleural effusion, lung collapse, or mediastinal or hilar fullness. • Chest CT scan shows size, location, and extent of primary tumour; evaluates for hilar and/or mediastinal lymphadenopathy and for distant metastases. • Sputum cytology shows malignant cells in sputum.

Condition	Differentiating signs / symptoms	Differentiating tests
		• Bronchoscopy shows endobronchial lesions. • Biopsy shows confirmation of malignancy.
Pulmonary tuberculosis	• Symptoms of cough, haemopytsis, and shortness of breath become progressive with corticosteroid treatment.	• HRCT shows upper lung non-layering cavitary lesions. • Sputum culture with positive AFB organisms.
Community-acquired pneumonia	• Symptoms of cough, sputum production, and fever subside with antibiotic therapy.	• Chest x-ray shows patchy infiltrates beginning to resolve with antibiotic therapy.
Bronchioloalveolar cell carcinoma	• Symptoms will be similar for bronchioloalveolar cell carcinoma and OP.	• There may be a pleural tag and the process bending or crossing the fissures on HRCT.
Coronavirus disease 2019 (COVID-19)	• Residence in/travel to a country/area or territory with local transmission, or close contact with a confirmed or probable case of COVID-19, in the 14 days prior to symptom onset. • Signs and symptoms are similar so it may be difficult to differentiate between the conditions clinically.	• Real-time reverse transcription polymerase chain reaction (RT-PCR): positive for severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) RNA. • It is not possible to differentiate COVID-19 from other causes of pneumonia on chest imaging.

Criteria

Epler and Colby pathological criteria[1] [2]

The pathological features of OP include:

- Organised polypoid granulation inflammatory tissue in the distal bronchiole airways, respiratory bronchioles, alveolar ducts, and alveoli
- Absence of extensive interstitial fibrosis
- Absence of traction bronchiectasis
- Absence of histological honeycombing.

Approach

OP is an inflammatory lung disease, so the general approach is to treat the inflammation. Corticosteroid therapy is the best option. The amount and length of treatment depend on the severity of disease and response to the medication. In some situations, OP can be monitored without treatment. Pulmonary rehabilitation and an exercise programme are helpful after the initial phase.

Severity of OP is determined by shortness of breath, extent of radiographic involvement, and pulmonary function tests, such as diffusing capacity <50% predicted and oxygen desaturation <85% during a walking exercise.

Rapidly progressive OP

Rapidly progressive OP begins with a flu-like illness proceeding to rapid progression of shortness of breath and respiratory failure within a few days. The patient is admitted to the ICU, supported with mechanical ventilation, and given intravenous corticosteroid therapy. This disorder may also be referred to as acute fibrinous organising pneumonia (AFOP).[56]

It is the author's experience that this type of OP is generally treated the same as acute interstitial pneumonia but requires high doses of corticosteroids and cyclophosphamide.

Cryptogenic OP

Corticosteroid therapy is the treatment of choice for cryptogenic OP. Oral prednisone (prednisolone) is prescribed at a higher dose (i.e., 40-60 mg/day) and tapered over 6 months.[57]

Severity is determined by symptoms of shortness of breath, extent of radiographic involvement, and diffusing capacity of <50% predicted. A total of 6 months of treatment may be effective for most patients, although 1 year may be required. About 5% percentage of patients may require intermittent doses for 3 to 5 years and this does not appear to affect mortality or morbidity.[57]

Mild disease may respond to macrolide antibiotics.[58] However, present studies are all observational and macrolides in cryptogenic OP need to be investigated in clinical trials. Erythromycin was used successfully in 6 Japanese women, with 1 responding at 2 months and 5 responding at 3 months.[59] Clarithromycin was also used successfully.[58] Azithromycin may also be effective. Macrolides may be useful for prevention of recurrence.

Secondary OP

Drug-related OP is reversible with drug cessation and/or corticosteroid therapy, depending on the severity of the disease. If symptoms are not severe, the causative drug could be stopped and any improvement. However, if symptoms are moderate or severe, the causative drug should be stopped and corticosteroids started immediately.

Toxin-exposure OP can be treated by immediately avoiding all contact with the toxin and prescribing corticosteroid treatment.

Post-radiation OP occurs in all regions of the lungs and will resolve without treatment or with corticosteroid therapy, depending on the severity of the OP.[22]

In post-infectious OP, the underlying infection either resolves on its own (some viral pneumonias) or is treated with appropriate antibiotics or antimalarials. Corticosteroid therapy is usually helpful and usually results in complete resolution.

When OP is associated with rheumatological or connective tissue disorders, it is often responsive to corticosteroid therapy.

Failure of corticosteroid treatment

If prednisone (prednisolone) is not effective or its dose cannot be weaned below 40 mg/day, azathioprine, and ciclosporin have been used with variable success as corticosteroid-sparing agents.[60]

In these situations, it is important to confirm that the primary process is OP (no honeycombing by high-resolution computed tomography scan) as many patients with seemingly corticosteroid-resistant OP do not have primary OP but have an underlying fibrosing process, such as interstitial pneumonia or non-specific interstitial pneumonia, not responsive to corticosteroid therapy. Here, OP is a secondary inflammatory lesion, responsive to corticosteroid therapy.

Treatment of recurrent OP

OP may recur in up to one third of patients. The symptoms will be the same as the initial episode, and the radiograph usually has the same pattern, although new lung regions may become involved.

If an OP recurrence has been established with recurrent symptoms, recurrence of radiographic findings, and deteriorating diffusing capacity, prednisone (prednisolone) is reinstituted at 20 mg higher than the dose at the time of recurrence. This new dose is given for 3 months, then tapered.[57] A second and third recurrence can be treated in the same way.

In very rare situations, lung transplantation may be necessary for patients who do not respond to treatment or have an unusual or hybrid form of OP.

Pulmonary rehabilitation

Rehabilitation is an important part of managing the mid-to-late phase of OP (after the initial few days of treatment, with the patient ambulatory and with improving symptoms and radiographic findings). It introduces an exercise programme for improving muscle conditioning, muscle oxygen efficiency, and sense of well-being. Patients also receive guidance for an ongoing exercise programme at home or at a commercial exercise facility.

Treatment algorithm overview

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Initial (summary)		
rapidly progressive OP		
	1st	**mechanical ventilation + intravenous corticosteroid followed by oral corticosteroid**
	plus	**cyclophosphamide**
	plus	**treatment of underlying causes/removal of causative factor**
	plus	**pulmonary rehabilitation**

Acute (summary)		
cryptogenic OP		
	1st	**oral corticosteroid**
	plus	**pulmonary rehabilitation**
	adjunct	**macrolide antibiotic**
	adjunct	**corticosteroid-sparing agent**
secondary OP		
	1st	**treatment of underlying cause/removal of causative factor**
	plus	**pulmonary rehabilitation**
	adjunct	**oral corticosteroid**
	adjunct	**corticosteroid-sparing agent**

Ongoing		(summary)
recurrent OP, rapidly progressive		
	1st	intravenous corticosteroid followed by oral corticosteroid
	plus	pulmonary rehabilitation
	adjunct	lung transplantation
recurrent OP, not rapidly progressive		
	1st	oral corticosteroid
	plus	pulmonary rehabilitation
	adjunct	lung transplantation

Treatment algorithm

Please note that formulations/routes and doses may differ between drug names and brands, drug formularies, or locations. Treatment recommendations are specific to patient groups: see disclaimer

Initial

rapidly progressive OP

1st **mechanical ventilation + intravenous corticosteroid followed by oral corticosteroid**

Primary options

» methylprednisolone: 250 mg intravenously every 6 hours for 3 days
-and-
» prednisolone: 2 mg/kg/day orally initially following methylprednisolone administration, slowly taper over weeks to months

» Rapidly progressive OP begins with a flu-like illness and progresses to shortness of breath and respiratory failure within a few days. The patient is admitted to the ICU and supported with mechanical ventilation.

» Intravenous methylprednisolone is given for 3 days, followed by oral prednisolone at a dose determined by the patient's weight. This dose is subsequently tapered over weeks to months, and patients should be discharged on oral corticosteroids.

plus **cyclophosphamide**

Treatment recommended for ALL patients in selected patient group

Primary options

» cyclophosphamide: consult specialist for guidance on dose

» It is the author's experience that this type of OP is generally treated the same as acute interstitial pneumonia but requires high doses of corticosteroids and cyclophosphamide.

plus **treatment of underlying causes/removal of causative factor**

Treatment recommended for ALL patients in selected patient group

» Drug-related OP is reversible with drug cessation.

» Toxin-exposure OP can be treated by avoidance of contact with the toxin.

Initial

» Post-radiation OP occurs in all regions of the lungs and will resolve without treatment.

» In post-infectious OP the infection either resolves on its own (some viral pneumonias) or is treated with antibiotics or antimalarials.

plus **pulmonary rehabilitation**

Treatment recommended for ALL patients in selected patient group

» This is an important part of managing the mid-to-late phase of OP (after the initial few days of treatment, with the patient ambulatory and with improving symptoms and radiographic findings). It introduces an exercise programme for improving muscle conditioning, muscle oxygen efficiency, and sense of well-being. Patients also receive guidance for an ongoing exercise programme at home or at a commercial exercise facility.

Acute

cryptogenic OP

1st **oral corticosteroid**

Primary options

» prednisolone: 40-60 mg orally once daily for 4 weeks, followed by 30-40 mg once daily for 4 weeks, followed by 20 mg once daily for 4 weeks, followed by 10 mg once daily for 6 weeks, followed by 5 mg once daily for 6 weeks

» Prednisolone remains the treatment of choice for OP.

» For most patients, 6 months of treatment is effective. In others, treatment may take 12 months.

» About 5% of patients require intermittent doses for 3 to 5 years and this does not appear to affect mortality or morbidity.[57]

plus **pulmonary rehabilitation**

Treatment recommended for ALL patients in selected patient group

» This is an important part of managing the mid-to-late phase of OP (after the initial few days of treatment, with the patient ambulatory and with improving symptoms and radiographic findings). It introduces an exercise programme for improving muscle conditioning, muscle oxygen efficiency, and sense of well-being. Patients also receive guidance for an ongoing exercise programme at home or at a commercial exercise facility.

adjunct **macrolide antibiotic**

Treatment recommended for SOME patients in selected patient group

Primary options

» erythromycin: consult specialist for guidance on dose

OR

» azithromycin: consult specialist for guidance on dose

OR

» clarithromycin: consult specialist for guidance on dose

MANAGEMENT

Acute

» Mild disease may respond to macrolide antibiotics.[58] However, present studies are all observational and macrolides in cryptogenic OP need to be investigated in clinical trials. Erythromycin was used successfully in 6 Japanese women, with 1 responding at 2 months and 5 responding at 3 months.[59] Clarithromycin was also used successfully.[58] Azithromycin may also be effective. Macrolides may be useful for prevention of recurrence.

adjunct **corticosteroid-sparing agent**

Treatment recommended for SOME patients in selected patient group

Primary options

» cyclophosphamide: consult specialist for guidance on dose

OR

» azathioprine: consult specialist for guidance on dose

OR

» ciclosporin: consult specialist for guidance on dose

» If prednisolone is not effective or its dose cannot be weaned below 40 mg/day, cyclophosphamide, azathioprine, and ciclosporin have been used with variable success as corticosteroid-sparing agents (with ongoing lower corticosteroid dose).[60]

» In these situations, it is important to confirm that the primary process is OP (no honeycombing by high-resolution computed tomography scan), as many patients with seemingly corticosteroid-resistant OP do not have primary OP but have an underlying fibrosing process, such as usual interstitial pneumonia or non-specific interstitial pneumonia, not responsive to corticosteroid therapy. Here, OP is a secondary inflammatory lesion, responsive to corticosteroid therapy.

secondary OP

1st **treatment of underlying cause/removal of causative factor**

» Drug-related OP is reversible with drug cessation.

Acute

» Toxin-exposure OP can be treated by avoidance of contact with the toxin.

» Post-radiation OP occurs in all regions of the lungs and will resolve without treatment.

» In post-infectious OP the infection either resolves on its own (some viral pneumonias) or is treated with antibiotics or antimalarials.

plus **pulmonary rehabilitation**

Treatment recommended for ALL patients in selected patient group

» This is an important part of managing the mid-to-late phase of OP (after the initial few days of treatment, with the patient ambulatory and with improving symptoms and radiographic findings). It introduces an exercise programme for improving muscle conditioning, muscle oxygen efficiency, and sense of well-being. Patients also receive guidance for an ongoing exercise programme at home or at a commercial exercise facility.

adjunct **oral corticosteroid**

Treatment recommended for SOME patients in selected patient group

Primary options

» prednisolone: 40-60 mg orally once daily for 4 weeks, followed by 30-40 mg once daily for 4 weeks, followed by 20 mg once daily for 4 weeks, followed by 10 mg once daily for 6 weeks, followed by 5 mg once daily for 6 weeks

» The addition of corticosteroids should form a part of treatment for toxin-exposure OP as well as severe cases of post-infectious OP and rapidly progressive OP. In drug-related and post-radiation OP not responsive to the removal of the causative agent, the addition of oral corticosteroids should also be considered. When OP is associated with rheumatological or connective tissue disorders, it is often responsive to corticosteroid therapy.

» For most patients, 6 months of treatment is effective. In others, treatment may take 12 months. About 5% of patients require intermittent doses for 3 to 5 years and this does not appear to affect mortality or morbidity.[57]

adjunct **corticosteroid-sparing agent**

Treatment recommended for SOME patients in selected patient group

Acute

Primary options

» cyclophosphamide: consult specialist for guidance on dose

OR

» azathioprine: consult specialist for guidance on dose

OR

» ciclosporin: consult specialist for guidance on dose

» If prednisolone is not effective or its dose cannot be weaned below 40 mg/day, cyclophosphamide, azathioprine, and ciclosporin have been used with variable success as corticosteroid-sparing agents (with ongoing lower corticosteroid dose).[60]

» In these situations, it is important to confirm that the primary process is OP (no honeycombing by high-resolution computed tomography scan) as many patients with seemingly corticosteroid-resistant OP do not have primary OP but have an underlying fibrosing process, such as usual interstitial pneumonia or non-specific interstitial pneumonia, not responsive to corticosteroid therapy. Here, OP is a secondary inflammatory lesion, responsive to corticosteroid therapy.

Ongoing

recurrent OP, rapidly progressive

1st **intravenous corticosteroid followed by oral corticosteroid**

Primary options

» methylprednisolone: 250 mg intravenously every 6 hours for 3 days, followed by oral prednisolone
-and-
» prednisolone: 2 mg/kg/day orally initially following methylprednisolone administration, slowly taper over weeks to months.

» OP may recur in up to one third of patients. The symptoms will be the same as the initial episode, and the radiograph usually has the same pattern, although new lung regions may become involved.

» If recurrent OP is rapidly progressing, the patient should be admitted to the ICU and supported with mechanical ventilation.

» Intravenous methylprednisolone is given for 3 days, followed by oral prednisolone at a dose determined by the patient's weight. This dose is subsequently tapered over weeks to months, and patients should be discharged on oral corticosteroid.

» If an OP recurrence has been established with recurrent symptoms, recurrence radiographic findings, and deteriorating diffusing capacity, following intravenous methylprednisolone, oral prednisolone can be reinstituted at 20 mg/day higher than the dose at the time of recurrence. This new dose can be given for 3 months, then tapered.[57]

» In those cases with a known cause, the underlying cause should be treated or causative factor removed.

» A second and third recurrence can be treated in the same way.

plus **pulmonary rehabilitation**

Treatment recommended for ALL patients in selected patient group

» This is an important part of managing the mid-to-late phase of OP (after the initial few days of treatment, with the patient ambulatory and with improving symptoms and radiographic findings). It introduces an exercise programme for improving muscle conditioning, muscle

Ongoing

oxygen efficiency, and sense of well-being. Patients also receive guidance for an ongoing exercise programme at home or at a commercial exercise facility.

adjunct **lung transplantation**

Treatment recommended for SOME patients in selected patient group

» Very rarely, lung transplantation may be necessary for patients who do not respond to treatment or who have an unusual or hybrid form of OP.

recurrent OP, not rapidly progressive

1st **oral corticosteroid**

Primary options

» prednisolone: 20 mg/day higher than dose at time of recurrence orally once daily for 12 weeks, then gradually taper according to response

» OP may recur in up to one third of patients. The symptoms will be the same as the initial episode, and the radiograph usually has the same pattern, although new lung regions may become involved.

» If an OP recurrence has been established with recurrent symptoms, recurrence radiographic findings, and deteriorating diffusing capacity, prednisolone can be reinstituted at 20 mg/day higher than the dose at the time of recurrence. This new dose can be given for 3 months, then tapered.[57]

» In those cases with a known cause, the underlying cause should be treated or causative factor removed.

» A second and third recurrence can be treated in the same way.

plus **pulmonary rehabilitation**

Treatment recommended for ALL patients in selected patient group

» This is an important part of managing the mid-to-late phase of OP (after the initial few days of treatment, with the patient ambulatory and with improving symptoms and radiographic findings). It introduces an exercise programme for improving muscle conditioning, muscle oxygen efficiency, and sense of well-being. Patients also receive guidance for an ongoing exercise programme at home or at a commercial exercise facility.

Ongoing

adjunct **lung transplantation**

Treatment recommended for SOME patients in selected patient group

» Very rarely, lung transplantation may be necessary for patients who do not respond to treatment or who have an unusual or hybrid form of OP.

Primary prevention

Occupational and environmental OP can be prevented by elimination of exposure to the agent, product substitution, ventilation, or work hazard preventive programmes.

Secondary prevention

For occupational and environmental OP, patients should be aware of the causative agent and appropriate preventive measures. For lung transplantation recipients, knowledge of symptoms of early recurrence is important, as early and active treatment of OP may prevent long-term disabling bronchiolitis obliterans and interstitial fibrosis.

Patient discussions

Patients should be provided with information about OP so that they will know what to expect from the condition and its treatment. The adverse reactions of corticosteroid therapy should be listed, explained, and understood. Patients should understand the symptoms of recurrent OP. Patients should also participate in a pulmonary rehabilitation programme and be assisted to develop an exercise programme.

Monitoring

Monitoring

Patients are initially monitored weekly, then monthly. Monitoring includes respiratory symptoms, chest x-rays, high-resolution chest computed tomography (HRCT) scans, and pulmonary function tests (including vital capacity and diffusing capacity, and oxygen desaturation during a walk in the hall or climbing stairs, or during the formal 6-minute walk test). A chest HRCT scan should be obtained for new-onset symptoms such as shortness of breath.

Complications

Complications	Timeframe	Likelihood
acute respiratory distress syndrome	**short term**	**high**
Can develop in patient with rapidly progressive OP. Signs and symptoms include dyspnoea and hypoxaemia, which can progress to acute respiratory failure.		
corticosteroid-related complications	**long term**	**high**
Include weight gain, cushingoid facies, friable ecchymotic skin, diabetes, hypertension, cataracts, osteoporosis, and aseptic necrosis of the hip.		

Prognosis

The overall mortality from OP is about 5%.[3] [6] [8] However, patients with rapidly progressive OP have a mortality of about 25%. Rapid diagnosis and initiation of corticosteroids are particularly critical in patients with rapidly progressive OP, to avoid death from a potentially treatable lesion.

Diagnostic guidelines

International

An official American Thoracic Society/European Respiratory Society statement: update of the international multidisciplinary classification of the idiopathic interstitial pneumonias (https://www.thoracic.org/statements/index.php)

Published by: American Thoracic Society; European Respiratory Society **Last published:** 2013

North America

ACR appropriateness criteria: chronic dyspnea-noncardiovascular origin (https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria)

Published by: American College of Radiology **Last published:** 2024

ACR appropriateness criteria: diffuse lung disease (https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria)

Published by: American College of Radiology **Last published:** 2021

Asia

Korean guidelines for diagnosis and management of interstitial lung diseases: part 4. cryptogenic organizing pneumonia (https://www.e-trd.org/journal/view.php?doi=10.4046/trd.2021.0025)

Published by: The Korean Academy of Tuberculosis and Respiratory Diseases **Last published:** 2021

Treatment guidelines

Asia

Korean guidelines for diagnosis and management of interstitial lung diseases: part 4. cryptogenic organizing pneumonia (https://www.e-trd.org/journal/view.php?doi=10.4046/trd.2021.0025)

Published by: The Korean Academy of Tuberculosis and Respiratory Diseases **Last published:** 2021

Key articles

- Epler GR, Colby TV, McLoud TC, et al. Bronchiolitis obliterans organizing pneumonia. N Engl J Med. 1985 Jan 17;312(3):152-8. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/3965933?tool=bestpractice.bmj.com)
- Epler GR. Bronchiolitis obliterans organizing pneumonia, 25 years: a variety of causes, but what are the treatment options? Expert Rev Respir Med. 2011 Jun;5(3):353-61. Abstract (http://www.ncbi.nlm.nih.gov/pubmed/21702658?tool=bestpractice.bmj.com)
- Bradley B, Branley HM, Egan JJ, et al; British Thoracic Society Interstitial Lung Disease Guideline Group, British Thoracic Society Standards of Care Committee; Thoracic Society of Australia; New Zealand Thoracic Society; Irish Thoracic Society. Interstitial lung disease guideline. Thorax. 2008 Sep;63 Suppl 5:v1-58. Full text (https://thorax.bmj.com/content/63/Suppl_5/v1.long) Abstract (http://www.ncbi.nlm.nih.gov/pubmed/18757459?tool=bestpractice.bmj.com)
- Travis WD, Costabel U, Hansell DM, et al. An official American Thoracic Society/European Respiratory Society statement: Update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med. 2013 Sep 15;188(6):733-48. Full text (https://www.atsjournals.org/doi/10.1164/rccm.201308-1483ST) Abstract (http://www.ncbi.nlm.nih.gov/pubmed/24032382?tool=bestpractice.bmj.com)
- Lazor R, Vandevenne A, Pelletier A, et al. Cryptogenic organizing pneumonia: characteristics of relapses in a series of 48 patients. Am J Respir Crit Care Med. 2000 Aug;162(2 Pt 1):571-7. Full text (https://www.atsjournals.org/doi/10.1164/ajrccm.162.2.9909015) Abstract (http://www.ncbi.nlm.nih.gov/pubmed/10934089?tool=bestpractice.bmj.com)

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Images

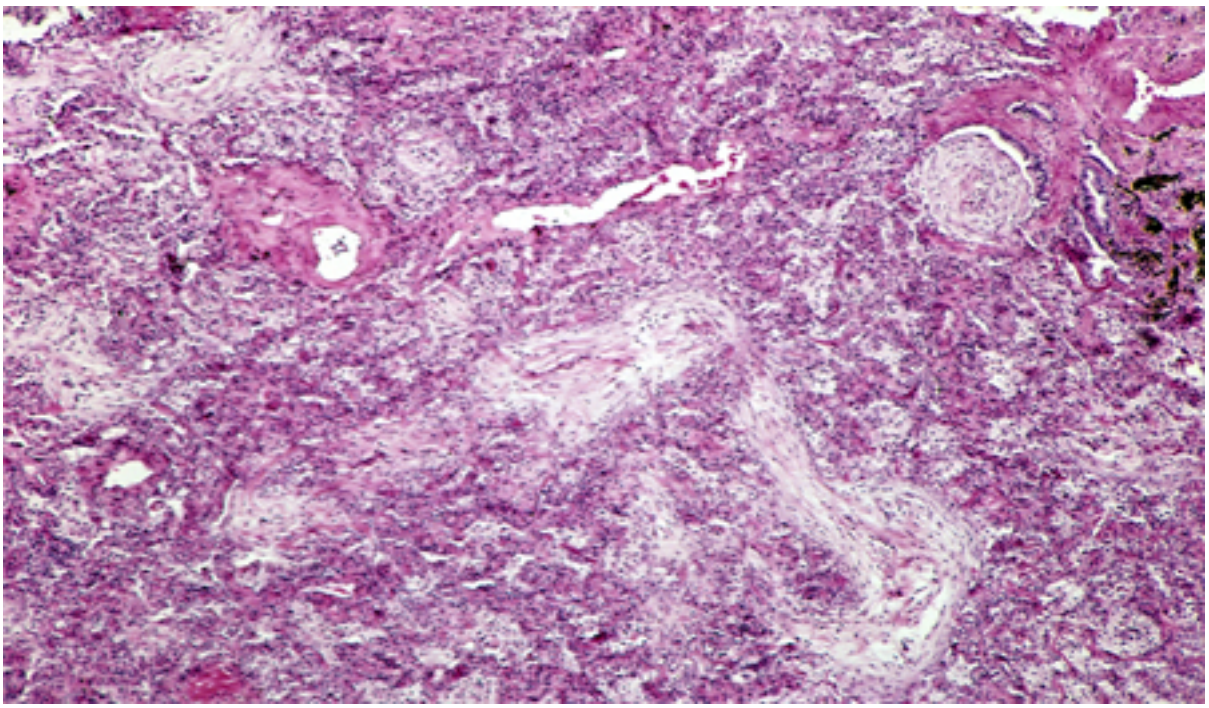

Figure 1: Medium-powered pathology slide showing circular and branching bronchioles filled with polypoid plugs of granulation tissue and alveoli filled with organising pneumonia

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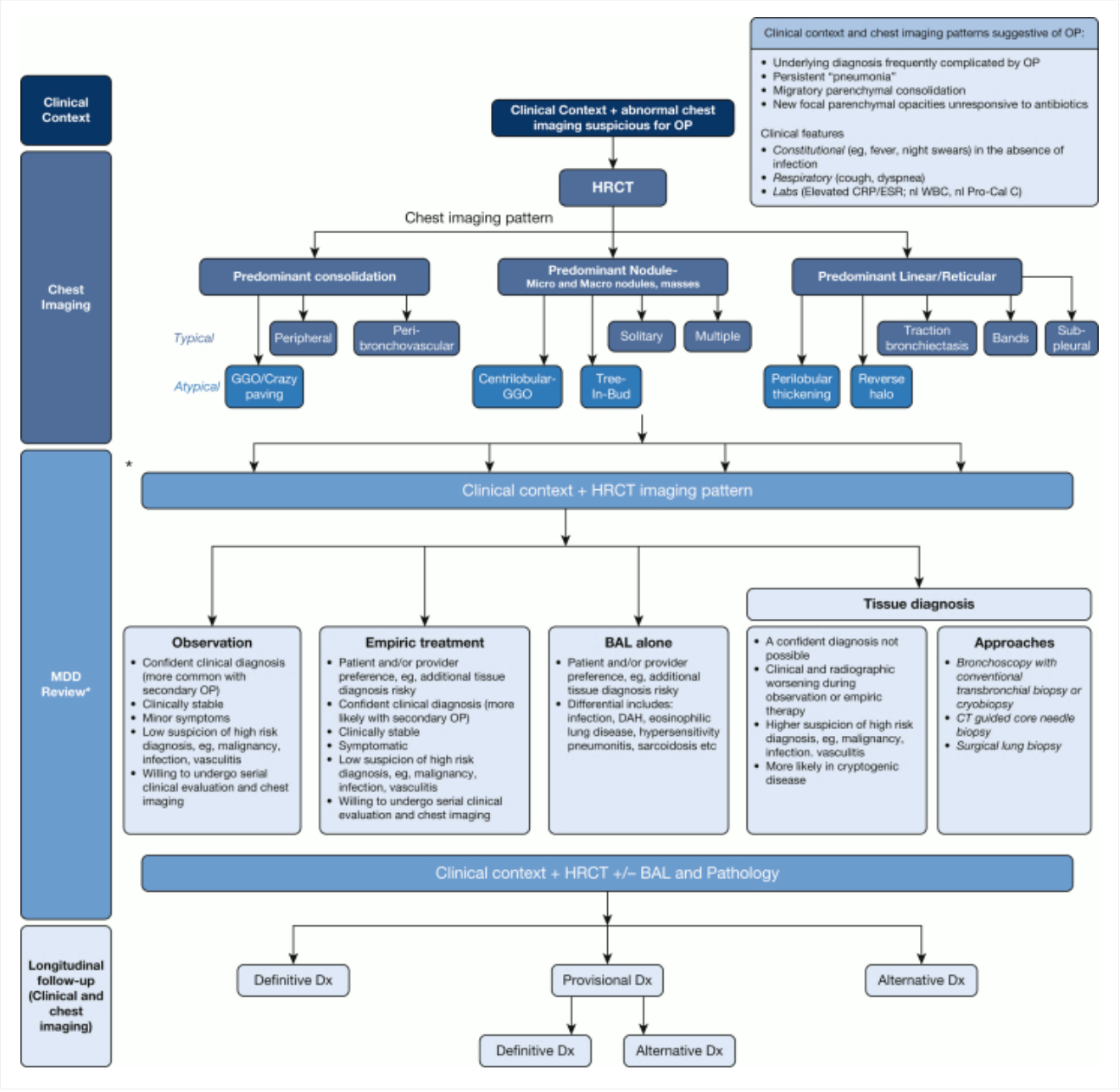

Figure 2: Algorithmic approach to organizing pneumonia. # A formal MDD may not be required in all cases, especially if the combination of clinical context and radiographic pattern is sufficiently convincing of the OP diagnosis. In such cases, a discussion between the physician and the radiologist is strongly encouraged. CRP = C-reactive protein; DAH = diffuse alveolar hemorrhage; Dx = diagnosis; ESR = erythrocyte sedimentation rate; GGO = ground-glass opacification; HRCT = high-resolution CT; MDD = multidisciplinary discussion; nl Pro-Cal C = normal procalcitonin; nl WBC = normal WBC; OP = organizing pneumonia

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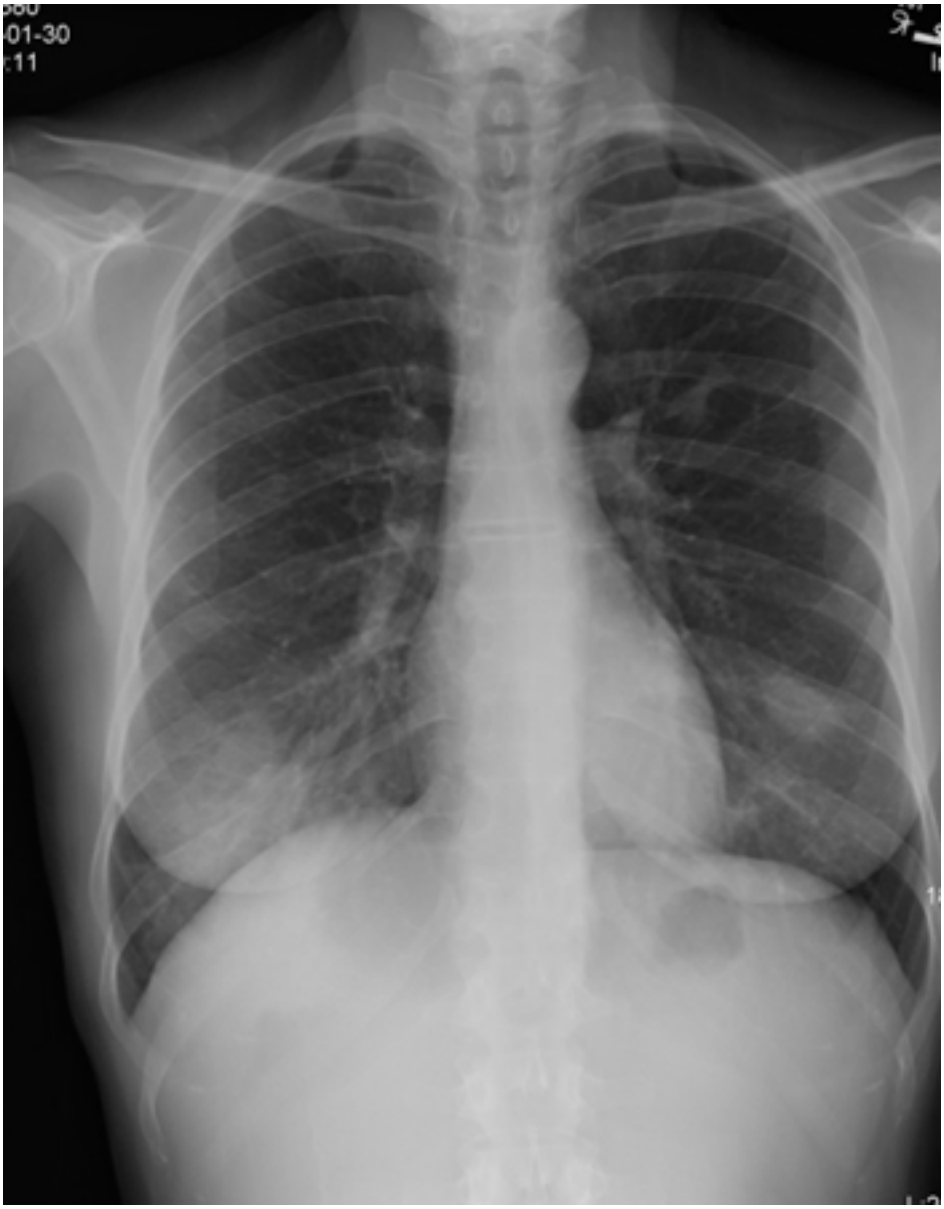

Figure 3: Chest x-ray showing bilateral patchy infiltrates

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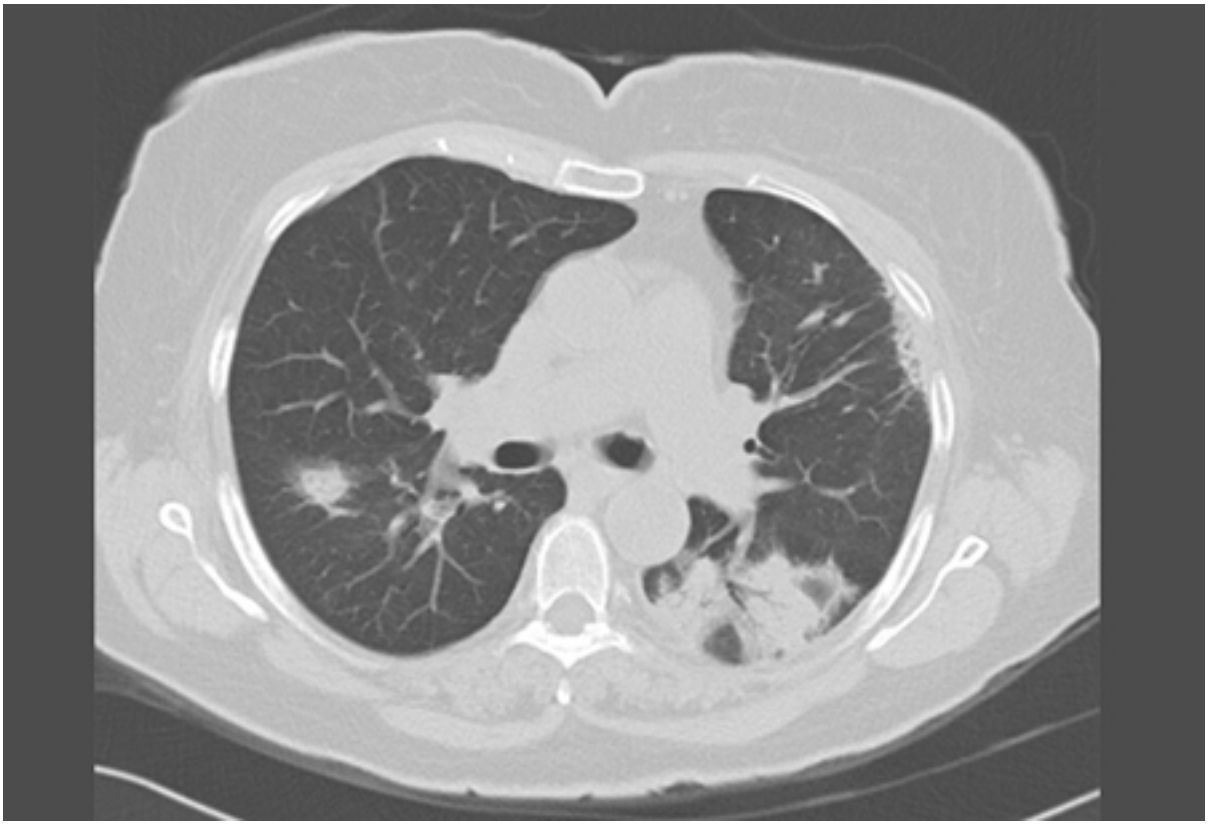

Figure 4: High-resolution chest CT showing bilateral ground glass opacities and a posterior triangular-based infiltrate with an air bronchogram

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Figure 1 – BMJ Best Practice Numeral Style

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