Cor Pulmonale

Synonyms: right-sided heart failure/right ventricular failure secondary to pulmonary disease

Description

Cor pulmonale describes impairment in right ventricular function as a result of respiratory disease, leading to increased resistance to blood flow in the pulmonary circulation.

The structure and function of the right ventricle is adversely affected by pulmonary arterial hypertension, induced by a disease process affecting the lungs, their ventilation or blood supply. For cor pulmonale to come about, mean pulmonary arterial pressure is usually >20 mm Hg. Complete right ventricular failure usually ensues if mean pulmonary arterial pressure is ≥40 mm Hg. It is thought that chronic hypoxia leads to pulmonary arteriolar constriction through excessive action of the physiological mechanism that acts to maintain the balance of ventilation and perfusion in the lungs.

Other mechanisms that may raise mean pulmonary arterial pressure in cases of cor pulmonale include:

- Chronic hypercapnia and respiratory acidosis causing pulmonary vasoconstriction.
- Anatomical disruption of the pulmonary vascular bed due to primary lung disease (for example, in emphysema, pulmonary thromboembolic disease and pulmonary fibrosis).
- Increased blood viscosity due to lung disease and its effects (for example, in secondary polycythaemia).

A wide range of pulmonary and cardiopulmonary disease processes may cause the condition. It is usually a chronic and progressive process; however, it does occur acutely due to sudden causes of pulmonary hypertension, usually following pulmonary embolism.

If right heart failure occurs due to primary disease of the left side of the heart, or because of a congenital cardiac lesion then it is not normally considered to be cor pulmonale.

Epidemiology

- There are few reliable figures for prevalence of cor pulmonale in the population at large, as the condition is difficult to diagnose reliably on the basis of clinical symptoms and signs alone.
- Its most common cause in the developed world is chronic obstructive pulmonary disease (COPD), due largely to tobacco smoking.
- Prevalence of cor pulmonale in COPD has been reported from 20-91% but it does not affect all those who have COPD\textsuperscript{[1]}.
- Acute cor pulmonale is most commonly due to massive venous thromboembolism and is a leading cause of death, the risk being highest within the first hour after the event.

Presentation

A diagnosis of cor pulmonale should be considered if any of the following is present: peripheral oedema, a raised venous pressure, a systolic parasternal heave or a loud pulmonary second heart sound. It is recommended by the National Institute of Health and Care Excellence (NICE) that the diagnosis of cor pulmonale be made clinically and that this process should involve excluding other causes of peripheral oedema\textsuperscript{[2]}. 
Symptoms
Common symptoms that may suggest the presence of cor pulmonale in a patient with pulmonary or cardiopulmonary disease include:

- Worsening tachypnoea (particularly at rest).
- Fatigue and lassitude.
- Ankle swelling.
- Worsening exertional dyspnoea (with deterioration in exercise tolerance).
- Worsening cough (particularly if non-productive).
- Angina-type chest discomfort - often non-responsive to nitrates (thought to be due to right ventricular ischaemia or stretching of pulmonary artery during exertion).
- Haemoptysis (due to pulmonary arteriolar rupture or leakage).
- Hoarseness - occurs occasionally (due to compression of the left recurrent laryngeal nerve by dilated pulmonary artery).
- Exertional syncope - a late symptom (indicating severe disease).
- Late-stage hepatic congestion can cause symptoms (anorexia, jaundice and right upper quadrant abdominal discomfort).

Signs

- Cyanosis and plethora.
- Chest markedly hyper-expanded.
- Laboured respiratory effort.
- Intercostal recession.
- Decreased air entry, and crackles and wheeze in the chest - due to underlying pulmonary pathology.
- Systolic bruits over lung fields - due to turbulent hyperdynamic pulmonary artery flow.
- Left parasternal or subxiphoid heave (a sign of right ventricular hypertrophy).
- Distended neck veins with raised and/or prominent JVP and visible a or v waves.
- 3rd/4th heart sounds and pansystolic murmur of tricuspid regurgitation over right heart.
- Split second heart sound with loud pulmonary component.
- Systolic ejection murmur with a sharp ejection click over the pulmonary artery (advanced sign).
- Diastolic pulmonary regurgitation murmur over the pulmonary artery (advanced sign).
- Marked hepatojugular reflux due to hepatic congestion.
- Hepatomegaly ± liver pulsatility if there is significant associated tricuspid regurgitation.
- Jaundice in advanced cases.
- Ascites in advanced cases.
- Peripheral pitting oedema.

Differential diagnosis

- **Primary pulmonary hypertension** (can be considered a cause of cor pulmonale).
- Pulmonary valve stenosis.
- Congestive cardiac failure due to primary cardiological disease.
- Congenital right-sided cardiac impairment.
- Right-sided heart failure due to right ventricular myocardial infarction.
- **Ventricular septal defect.**
Investigations

Investigation of underlying cardiopulmonary disease
The following investigations are often used to delineate the cause(s) of respiratory compromise that may lead to cor pulmonale and to inform optimal management:

- Alpha-1 antitrypsin levels if considered relevant.
- Autoantibody screen if there is suspected collagen vascular disease.
- Thrombophilia screen if there is suspected chronic venous thromboembolism (proteins C and S, antithrombin III, factor V Leiden, antithromboplastin antibodies, homocysteine levels).
- CXR (allows assessment of right atrial size and pulmonary artery enlargement).
- Spirometry/lung function tests including gas transfer and flow volume loop.
- CT/MRI scan of the chest.
- Bronchoscopy.
- Lung biopsy (open or transbrachial).
- Ventilation/perfusion scan/spiral-CT angiography/MRI-angiography (where there is reason to suspect recurrent pulmonary embolism or acute right heart failure due to thromboembolic disease).

Investigation of right heart function and cardiopulmonary function

- ECG (looking for evidence of right ventricular hypertrophy and strain/dysrhythmias associated with impaired right ventricular function).
- FBC to determine haematocrit where there is secondary polycythaemia.
- Arterial/capillary blood gases on room air and in response to administration of oxygen.
- Brain natriuretic peptide (BNP) assay (elevated BNP levels have been shown to correlate with raised pulmonary artery pressures and presence of cor pulmonale).  
- Continuous-wave Doppler echocardiography allows right ventricular systolic pressure to be calculated.
- Pulsed-wave Doppler echocardiography - allows estimation of pulmonary artery systolic pressure.
- Two-dimensional echocardiography - assesses right ventricular size.
- Radionuclide ventriculography - measures right ventricular ejection fraction.
- Ultra-fast ECG-gated CT scanning - currently used experimentally to assess right ventricular function but may become more widely used.
- Right heart catheterisation - an invasive test that may be poorly tolerated in patients with very poor cardiorespiratory reserve; gives accurate measured, rather than estimated, values.

Causative diseases

Those due to secondary pulmonary arterial compromise

- COPD (by far the most common).
- Other causes of parenchymal lung disease - eg, idiopathic fibrosing alveolitis, emphysema, pneumoconiosis, cystic fibrosis.
- Neuromuscular disorders causing chronic hypventilation - eg, polio, myasthenia gravis, motor neurone disease.
- Obstructive or central sleep apnoea/Pickwickian syndrome (obesity hypoventilation syndrome).
- Thoracic deformity - eg, kyphoscoliosis.
- Atelecic or capillary dysplasia.
- Neonatal pulmonary disease and its sequelae - eg, bronchopulmonary dysplasia.

Those due to primary disease of the pulmonary arterial vessels

- Recurrent pulmonary emboli.
- Other pulmonary veno-occlusive disease.
- Pulmonary vasculitis.
- Sickle cell disease.
- Altitude sickness/pulmonary vasoconstriction due to chronic altitude exposure.
- Primary pulmonary hypertension.
Management

Patients presenting with cor pulmonale should be assessed for the need for long-term oxygen therapy. Oedema associated with cor pulmonale can usually be controlled symptomatically with diuretic therapy. The following are not recommended for the treatment of cor pulmonale: angiotensin-converting enzyme inhibitors, calcium-channel blockers, alpha-blockers or digoxin (unless there is atrial fibrillation) [2].

Acute cor pulmonale is treated by trying to rapidly correct the underlying precipitant, which is often acute pulmonary embolism or an infective exacerbation of COPD. Standard treatment for these conditions is used in an attempt to correct the underlying cause of acute right heart failure.

Similarly, in chronic cor pulmonale, treatment of the underlying cause is combined with specific management as below:

- **Long-term oxygen therapy (LTOT)/nocturnal oxygen therapy (NOT)** have been shown to improve quality of life and survival in patients with severe chronic hypoxia due to lung disease, by reducing pulmonary arteriolar constriction and improving/slowing the progression of cor pulmonale [2]. They are usually recommended where $P_aO_2$ is <55 mm Hg or $S_aO_2$ is <88%. A Cochrane review has confirmed these benefits but shown a lack of efficacy for patients with only mild-to-moderate hypoxaemia/patients that only desaturate at night [5]. Where there is clear clinical/investigational evidence of cor pulmonale, and higher mental/cognitive impairment attributable to hypoxia complicating chronic lung disease, LTOT/NOT may be given with oxygenation above these values. Great care must be taken to ensure the safety of patients who continue to smoke, as oxygen is highly combustible, and many clinicians will not give oxygen therapy to smokers (another reason being negation of its benefit by the presence of elevated carboxyhaemoglobin levels in smokers).

- **Diuretics** such as furosemide and bumetanide are frequently utilised (particularly where the right ventricular filling volume is markedly elevated) and in the management of associated peripheral oedema. Care must be taken to avoid over-diuresis which can impair the function of both ventricles. It may also induce a hypokalaemic metabolic alkalosis which can lessen respiratory drive through reducing the hypercapnic stimulus to breathe. Intravenous diuretics may be needed in patients with acute decompensation and severe peripheral oedema, due to poor absorption of oral medication from the oedematous gut.

- **Vasodilators** such as nifedipine and diltiazem have been shown to have modest physiological effects, although there is no convincing trial evidence of their efficacy [6].

- **Inotropic drugs**, particularly digoxin, are frequently used but there is little evidence for their efficacy in right heart failure, in contrast to their use with left ventricular failure [7].

- **Methylxanthine bronchodilators** such as theophylline are frequently used for their beneficial effect on bronchial tone and concomitant mild positive inotropic effect [8].

- **Anticoagulation** is used where patients have venous thromboembolism as the underlying cause of their cor pulmonale, and where there are significant risk factors for venous thromboembolic disease in patients with chronic lung disease and cor pulmonale. There is little evidence of tangible benefit in terms of survival in cases due to secondary pulmonary hypertension, in contrast to their proven benefit in primary pulmonary hypertension.

- **Venesection** is used with caution in some patients who have severe secondary polycythaemia (usually defined as haematocrit >0.65) due to chronic hypoxia. It has been shown to improve symptomatology; however, there is no evidence of improved survival.

- **Transplantation** of single/double lung or heart/lung is used in some extreme cases of cor pulmonale and significantly improves outlook. The underlying cause must usually be unrelated to smoking to reduce the likelihood of other pathology that would give poorer outcomes.

Complications

- Exertional syncope.
- Hypoxia and significantly limited exercise tolerance.
- Peripheral oedema.
- Peripheral venous insufficiency.
- Tricuspid regurgitation.
- Hepatic congestion and cardiac cirrhosis.
- Death.

Prognosis

This is dependent on the nature of the underlying cause and its rate of progression. The overall five-year survival rate for cor pulmonale complicating COPD is approximately 50% [6].

Long-term oxygen therapy improves this and the best prognostic indicator is the pulmonary arterial pressure. Prognosis also appears to be significantly improved by smoking cessation and correct use of LTOT/NOT [6].

Prevention

Progression of cor pulmonale can be slowed by strict adherence to smoking cessation and appropriate use of LTOT/NOT. COPD-related cor pulmonale is preventable by not starting to smoke, or stopping smoking before COPD becomes a significant clinical problem.

Further reading & references

- Chronic obstructive pulmonary disease; NICE CKS, September 2015 (UK access only)

2. Chronic obstructive pulmonary disease; NICE Clinical Guideline (2010)

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