Continuing professional development
Pulmonary Vascular Diseases

Module 1. Anatomy and development including malformations

1. Pulmonary vasculature and lymphatic drainage
2. Anatomy of the cardiovascular system
3. Right ventricular anatomy

Module 2. Immunology and defence mechanisms

1. Immunology

Module 3. Ventilation

1. Physiology of tidal breathing
2. Active inspiration and passive expiration
3. Relative elastic properties of the lung and chest wall
4. Understanding lung volumes
5. Transpulmonary pressures and breathing
6. Changes in intrathoracic pressure
7. Disease-specific effects on ventilation
8. COPD and interstitial lung disease (ILD)

Module 4. Circulation

1. Normal pulmonary vascular pressures and flows
2. Effects of exercise
3. Active hypoxic and neurohumoral regulation of pulmonary circulation
4. Pulsatile flow pulmonary haemodynamics
5. Non-invasive evaluation of pulmonary circulation
6. Invasive evaluation of pulmonary circulation
7. Interpretation of pulmonary vascular resistance
8. Components of pulmonary vascular load, right ventricular function (systolic and diastolic)

Module 5. Gas exchange

1. Transfer factor of the lung for carbon monoxide (TLCO)
2. Definition
3. Technique
4. Calculation of TLCO and measurement of carbon monoxide transfer coefficient (KCO)
5. Implications of KCO × alveolar volume (VA) = TLCO

Module 6. Diagnostics – exercise protocols

1. Field tests *versus* ergometer/treadmill tests
2. Maximal incremental test
3. Walking tests: 6-minute walking distance (6MWD) and incremental shuttle walk test (ISWT)
4. Indications for cardiopulmonary exercise testing (CPET)
5. Exercise variables and indexes
6. Maximal $V'O_2$, heart rate (HR) and ventilation
7. Lactate threshold/ventilatory thresholds
8. Oxygen pulse-HR reserve and maximum predicted HR: $V'E$-$V'CO_2$ slope and ventilatory equivalent
9. Carbon dioxide-breathing reserve
10. Dynamic hyperinflation
11. Arterial oxygen desaturation
12. Tolerable limit of exercise and “isotime” measurements
13. CPET response patterns
14. Ventilatory response
15. Pulmonary gas exchange
16. Cardiovascular response

17. Exercise testing in prognostic evaluation
18. Evaluation of the effects of therapeutic interventions
19. Somnography/polygraphy
20. Activity monitoring
21. Evaluation of psychological and social status: SYMPACT and EMPHASIS10 questionnaires

Module 7. Signs and symptoms suggesting pulmonary vascular disease (PVD)

1. Evidence of right heart failure
2. Dyspnoea
3. Reducing exercise tolerance
4. Presyncope
5. Syncope
6. Chest tightness

Module 8. Disease processes associated with PVD

1. Connective tissue disease (CTD)
2. Congenital heart disease
3. Hepatic disease
4. HIV
5. ILD including sarcoidosis
6. COPD including emphysema
7. Lung hypoventilation
8. Pulmonary embolic disease
9. Left heart failure
10. Genetic causes of PVD (*i.e.* BMPR2, EIF2AK4 and HHT genes)
11. Haematological diseases (*e.g.* polycythaemia rubra vera)
12. Portal hypertension

Module 9. Identification of signs of PVD by imaging modalities

1. Acute pulmonary embolism
2. Chronic thromboembolic disease
3. Evidence of congenital heart disease
4. Abnormal pulmonary venous return
5. Enlarged pulmonary arteries
6. Dilated right heart
7. Right ventricular hypertrophy
8. Mosaicism
9. Pulmonary arteriovenous malformations
10. Pulmonary vein stenosis

Module 10. Right heart catheterisation

1. Use of right heart catheterisation
2. Definition of pulmonary hypertension (PH)
3. Indications, physiologic considerations and interpretation of exercise haemodynamic testing

Module 11. PH-specific therapies

1. Therapy
2. Different types of delivery
3. Medical management of right heart failure
4. Non-medical therapies
5. Surgery/balloon pulmonary angioplasty for chronic thromboembolic pulmonary hypertension (CTEPH)
6. Pulmonary artery denervation

Module 12. Pulmonary rehabilitation

1. Indications for pulmonary rehabilitation

Module 13. Palliative care

1. Indications for institution of palliative care
2. End-of-life relief of symptoms by:
3. Pharmacological interventions
4. High-flow nasal oxygen
5. Discussing end-of-life decisions with patients and their relatives

Module 14. Oxygen therapy

Module 15. Mechanically assisted ventilation

1. Indications for assisted ventilation
2. Effects of mechanical ventilation on pulmonary haemodynamics and heart function

Module 16. Indication for lung transplantation

1. Principles of evaluation for lung transplantation
2. Risk and particularities of lung transplantation in patients with PVD
3. Severity grading of lung diseases with respect to referring a patient to a transplant centre
4. Extracorporeal membrane oxygenation/right ventricular assist device

Module 17. Evaluation of respiratory emergencies

1. Management of right heart failure
2. Management of cardiac arrhythmias
3. Management of acute pulmonary embolism (PE), pulmonary embolism response team (PERT)
4. Cardiopulmonary arrest in a patient with pulmonary arterial hypertension (PAH)

Module 18. Differential diagnosis, immediate management steps and specific conditions

1. Differential diagnosis of PVD and PE
2. Haemoptysis
3. Dyspnoea
4. Thoracic pain
5. PE
6. Haemoptysis
7. Dyspnoea
8. Thoracic pain
9. Massive acute pulmonary embolism
10. Management of shock related to right heart failure
11. Early identification of chronic thromboembolism

Module 19. COPD and emphysema

1. Severe PH sub-phenotypes in COPD
2. Polycythaemia
3. Management of cor pulmonale

Module 20. Obstructive sleep apnoea syndrome

1. Signs and symptoms associated with sleep-disordered breathing
2. Recognition of sleep-disorder breathing as a cause of moderate PH
3. Physiology and pathophysiology of sleep apnoea syndromes relevant to acute respiratory failure

Module 21. Sarcoidosis

1. Recognition of Sarcoidosis and its association with PH
2. Recognition of PH as a poor prognostic factor in sarcoidosis
3. Effects of drug therapy on PH

Module 22. ILD

1. Prognostic factors associated with ILD and PVD
2. Classification of ILD
3. Limitations/dangers of PAH therapies in ILD

Module 23. CTD related to ILD

1. ILD associated with CTD
2. Prognostic factors associated with ILD

3. List treatment choices

Module 24. Drug-induced disease

1. Identification of causes of PVD from drug history
   1.1. Anorexigens
   1.2. Amphetamines
   1.3. Selected tyrosine kinase inhibitors
   1.4. Interferon
   1.5. Chemotherapeutic agents (alkylating agents)

Module 25. Thromboembolic disease

1. Identification of PE
2. Differential diagnosis of PE
3. Aetiology of PE
4. Pulmonary Embolism Severity Index score and risk stratification of PE
5. Management of PE without acute hemodynamic compromise
6. Conditions/diseases associated with an increased risk of thromboembolic disease
7. Current indications for prophylaxis against thromboembolic disease
8. Adverse effects associated with anticoagulation therapy
9. Interpretation of blood cell counts and coagulation laboratory tests
10. Drug indications and their appropriate dosages
11. Willingness to consider potential candidates for prophylaxis against thromboembolic disease
12. Prophylaxis against thromboembolic disease
13. Conditions/diseases associated with an increased risk of thromboembolic disease
14. Adverse effects associated with anticoagulation therapy
15. Interpretation of coagulation laboratory tests
16. Drug indications and their appropriate dosages
17. Genetic risk factors for thrombosis and their impact on clinical decisions
18. Indications for targeted thrombosis (interventional radiology) and embolectomies
19. Diagnosis and management of CTEPH
20. Indications for thrombophilia screening
21. Indications for inferior vena cava filter placement
22. Follow-up after PE presentation, *e.g.* risk scoring (such as DASH) to discuss the risks/benefits of anticoagulation therapy
23. Pancreatic enzyme replacement therapy

Module 26. PH

1. Pathophysiology of PH
2. Pharmacological treatment of PH according to underlying disease
3. Diagnosis of PH and cor pulmonale
4. Risk stratification
5. Translating national and international management guidelines to an individual patient
6. Appropriate decisions for referral and transfer to specialised referral centres
7. Prognosis of patients with PH in acute care settings
8. Indications for systemic pharmacotherapy
9. Indications for combination therapy
10. Indications for intravenous therapy
11. Chronic monotherapy or combination systemic therapy
12. Institute general therapeutic and supportive measures

Module 27. Vasculitis and diffuse pulmonary haemorrhage

1. Pulmonary haemorrhage and haemoptysis
2. Differential diagnoses, diagnostic steps and therapeutic options for pulmonary haemorrhage and haemoptysis

3. Triage by severity
4. Indications for radiological evaluation
5. Indications for bronchoscopic evaluation

Module 28. Arteriovenous (AV) malformation

1. Symptoms of AV malformation
2. Diseases associated with AV malformation (*e.g.* hereditary haemorrhagic telangiectasia)
3. Diagnostic strategies when AV malformation is suspected
4. Therapeutic interventions

Module 29. Cardiac disease

1. Differentiate between cardiac and pulmonary disease as a cause of acute respiratory failure
2. Recognition of cardiac disease as an aetiology of PH
3. Monitoring cardiac dysfunction in PVD patients via clinical, laboratorial, functional, echocardiographic and radiological means
4. Principles of invasive cardiovascular monitoring (*e.g.* Swan-Ganz catheterisation)
5. Cardiovascular effect of positive pressure ventilation
6. Congenital heart diseases and Eisenmenger syndrome

Module 30. Gastrointestinal, liver and kidney disease

1. Portopulmonary hypertension
2. Hepatopulmonary syndrome

Module 31. Haematological disease

1. PVD in patients with past or current haematological conditions
2. Thrombophilia as a potential sign of PVD

Module 32. Connective Tissue Diseases (CTD)

1. CTD as a cause of PH
2. Symptoms and clinical presentation
3. Pathophysiology of CTD and PH
4. Differential diagnosis and optimal testing
5. Optimal treatment
6. Prognostication of identified CTD

Module 33. Hereditary PAH

1. Familial occurrence of PH

Module 34. High-altitude sickness

1. High-altitude as a cause of pulmonary oedema (HAPE)
2. Pathophysiology, symptoms and clinical presentation of high-altitude sickness
3. Performance of tests to establish the diagnosis
4. Preventive measures