

Expert Group Pack C: Pulmonary Hypertension

STATION C - PULMONARY HYPERTENSION

Read this pack with your group. Each person takes one section to specialise in and teach to visitors.

What is Pulmonary Hypertension?

Abnormally high blood pressure in the arteries that supply the lungs. Defined as a resting mean pulmonary artery pressure ≥ 20 mmHg, measured by right heart catheterisation.

KEY DISTINCTION: This is NOT the same as the hypertension you studied last week. Systemic hypertension = high blood pressure in your body's arteries (measured with an arm cuff). Pulmonary hypertension = high blood pressure specifically in the arteries between your heart and lungs. You can have one without the other.

Five WHO groups based on cause:

Group 1: Pulmonary arterial hypertension (PAH) - disease of the small pulmonary arteries themselves

Group 2: PH due to left-sided heart disease (the most common overall)

Group 3: PH due to lung disease (e.g. COPD, ILD) - very relevant to today's session

Group 4: Chronic thromboembolic PH (blood clots in the pulmonary arteries)

Group 5: PH with unclear or multifactorial mechanisms

Prevalence

Global: Estimated ~1% of the population, rising to ~10% in people over 65.

UK: PAH specifically: incidence ~1.1-2.4 per million per year; ~8,000 new referrals to PH centres per year.

PH in COPD: Develops in ~38-50% of patients with severe COPD - a common complication of a very common condition.

Demographics: More common in women than men (ratio ~1.8:1). Median age at diagnosis ~60. More than 25% diagnosed over 70.

Diagnostic delay: Average 2-3 years from first symptoms to diagnosis. Frequently misdiagnosed as asthma, anxiety, or deconditioning.

Aetiology & Pathology

The core problem: The walls of small pulmonary arteries thicken and stiffen through four processes:

1. Vasoconstriction - the arteries tighten
2. Vascular remodelling - smooth muscle and the inner lining (intima) thicken permanently
3. In situ thrombosis - small blood clots form within the pulmonary arteries
4. Inflammation - inflammatory cells infiltrate the vessel walls

The consequence: The RIGHT ventricle has to pump harder and harder to push blood through these narrowed arteries. Initially it compensates by thickening (hypertrophy). Eventually it weakens, dilates, and fails. Right ventricular failure is the primary cause of death in PH.

Three dysfunctional pathways (these are the drug targets):

- Reduced nitric oxide (NO) → less vasodilation
- Reduced prostacyclin → less vasodilation + increased platelet aggregation
- Increased endothelin-1 → more vasoconstriction + smooth muscle proliferation

Signs and Symptoms

- Progressive breathlessness on exertion (the most common presenting symptom)
- Fatigue - disproportionate to the level of activity
- Dizziness or syncope (fainting) - particularly on exertion
- Chest pain - from right ventricular strain
- Peripheral oedema - swollen ankles and legs (right heart failure)
- Ascites - fluid accumulation in the abdomen (advanced disease)
- Cyanosis - blue lips and extremities

Diagnostic challenge: All of these symptoms are non-specific and overlap with many other conditions. The average delay from first symptoms to diagnosis is 2-3 years. PH is frequently misdiagnosed as asthma, anxiety, or simple deconditioning.

Risk Factors

- Existing lung disease (COPD, ILD, pulmonary fibrosis) - chronic hypoxia drives vascular remodelling
- Left-sided heart disease (heart failure, valvular disease)
- Connective tissue diseases (scleroderma is one of the strongest risk factors for PAH)
- Blood clots (chronic thromboembolic disease)
- HIV infection
- Family history / genetic mutations (BMP2 gene)
- Female sex (for PAH specifically)
- Obesity, smoking, ageing (for Group 3 PH)

Impact on Quality of Life

- 83% of patients report difficulty climbing stairs; 97% report affected sports/exercise participation
- Severe limitations on daily activities: housework, shopping, cooking, personal care in advanced stages
- Employment: many reduce hours or stop working entirely
- Social isolation from inability to participate in physical activities
- Psychological: depression, anxiety, fear of fainting in public, grief for lost independence
- Financial: specialist medications are extremely expensive; need to travel to specialist PH centres
- Prognosis: untreated PAH has median survival ~2.8 years. Modern treatment improves this, but it remains life-limiting

WALES SPOTLIGHT

PH connects directly to last week: it is a complication of heart failure AND a consequence of COPD - both highly prevalent in Wales.

Wales' high COPD prevalence means a correspondingly high burden of Group 3 PH.

The heart-lung connection is central: when the lungs are diseased, the heart pays the price. The right ventricle pumps to the lungs - diseased lungs mean increased resistance - the heart fails.

PH illustrates why we can't study organ systems in isolation - cardiovascular and pulmonary disease are deeply intertwined.