

A shared vision for pulmonary arterial hypertension in the UK

A policy consensus statement



What is this policy consensus statement?

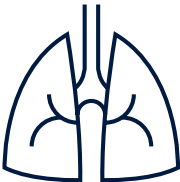
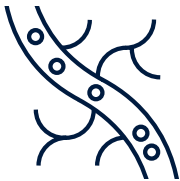


This statement outlines a shared vision for the future of pulmonary arterial hypertension (PAH) care in the UK, with a focus on reducing time to accurate diagnosis – an issue which is consistently flagged by people living with the condition.

This policy consensus statement has been developed and fully funded by MSD. Insights were gathered during an MSD-organised multidisciplinary workshop (November 2023) and an MSD-sponsored session at the PH Professionals Spring Meeting (February 2024), both fully funded by MSD, as well as additional feedback gained from experts via email.



What is pulmonary arterial hypertension?



At a glance

- PAH is a chronic and incurable disease that causes the arterial walls in the lungs to thicken.¹ Pressure within the arteries increases and, over time, this can lead to right heart failure and premature death.²
- PAH is a rare sub-group of a broader condition called pulmonary hypertension (PH).³
- In March 2023, 3,816 people in the UK had an active referral to a specialist service for PAH (out of 8,777 referrals for PH).⁴
- PAH affects more females than males, with UK and European registries suggesting a ratio of up to 1.6 to 1.⁵
- A PH diagnosis is a life-changing event, for people living with the condition and their loved ones. In a survey by the Pulmonary Hypertension Association UK, 84% of respondents reported that PH had an impact on their mental or emotional wellbeing.⁶
- The main symptom for PAH is breathlessness,⁷ but other symptoms may include dizziness, feeling faint, swelling of the feet or ankles and chest pain (particularly during exercise).⁸
- The survival rate for PAH is approximately 50% of adult patients at five years post-diagnosis.⁴ However, with appropriate management, many people can enjoy fulfilling lives with some lifestyle adaptations.⁹

3,816

people in the UK had an active referral for PAH in 2023

84%

of PH patients report an impact on their mental health and wellbeing

50%

of adults with PAH are alive 5 years post-diagnosis



PAH is a rare and debilitating disease that causes the arteries between the heart and lungs to constrict and narrow.¹ Pressure within the arteries increases and, over time, this can lead to right heart failure and premature death.² For adults with PAH, the survival rate is approximately 50% in the five years after diagnosis.⁴ In a national patient survey by the Pulmonary Hypertension Association UK (PHA UK), 50% of respondents stated that PH has a ‘major impact’ on their lives.⁶ However, with timely management, many people can have a good quality of life with some lifestyle adaptations.⁹

“A lot of people don’t understand what it means to have PH and how it can affect your life.”

- Breathless, not voiceless survey (PHA UK, 2023)

Detection and time to diagnosis of PAH remains a persistent challenge in the UK. This is partly due to its rarity and non-specific symptoms (e.g. breathlessness, fatigue), which might be misattributed to anxiety or other more common respiratory diseases.¹⁰ In a PHA UK survey, 51% of respondents reported waiting more than a year for a PH diagnosis after first noticing symptoms. For a quarter of respondents, this wait took longer than three years.⁶ More than a third of respondents were first diagnosed during an emergency hospital admission.¹¹

Did you know?

Delayed diagnosis not only worsens outcomes and limits treatment options for patients,¹² but it also generates considerable costs for the NHS through repeat consultations and hospital admissions.¹³

“I had symptoms for a good couple of years. Doctors thought it was asthma and went through a long process of changing different inhalers, using steroids, and nothing really improving.”

- Breathless not voiceless survey (PHA UK, 2023)

Diagnosis and care are led by specialist PH centres, which are unevenly geographically distributed across the UK.¹⁴ Patient feedback on specialist-led care is positive and specialist centres consistently meet national quality standards – a PHA UK survey found that 91% of respondents describe their care at specialist centres as excellent or good.^{2,6} While innovations in treatment have helped to relieve symptoms and improve quality of life,² there is much more we can do to ensure patients reach specialist centres sooner for timely diagnosis and care.

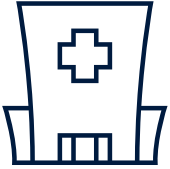
Did you know?

There are seven specialist PH centres in England (four in London) and one in Scotland¹⁴ Shared Care Centres have also been set up in England and Northern Ireland, with formal links to specialist centres to provide ongoing care and management of PH.¹⁵

92% of respondents in the PHA UK survey said it is better to travel to a specialist centre than to be under the care of a non-PH specialist at a more local hospital.



How does PH impact people, the NHS and the UK economy?



Beyond the personal impact, PH (including PAH) generates considerable costs for the NHS through repeat consultations and hospital admissions,¹³ and to the wider economy through loss of workforce productivity.

- **Repeat appointments:** In a PHA UK survey, 32% of respondents reported seeing four or more doctors before receiving their PH diagnosis.⁶ Based on this figure, we calculated an estimated cost to the NHS of at least £442,406 (based on the average cost of a general practice appointment of £30¹⁶ and 11,521 managed patients recorded in the 14th NHS National PH Audit⁴).
- **Hospital care:** PAH cost the NHS in England an estimated £43.2 million between 2013 and 2017 (£33.9 million in admissions, £8.3 million in outpatient visits and close to £1 million in A&E attendance).¹³
- **Labour hours lost:** In a PHA UK survey, 44.4% of respondents were not working at the time of diagnosis. Of those who were working, 28% of respondents reported giving up work completely and 23% went on long-term sick leave.¹⁷ Based on this research, we have calculated an estimated loss of 57,193 labour hours per year amongst people working at the time of diagnosis (does not include the impact of reduced hours, which was reported by 22% of respondents working at the time of diagnosis).

£442k

estimated cost to the NHS for general practitioner appointments before diagnosis

£43.2m

estimated PAH cost to NHS between 2013 and 2017

57,193

labour hours lost per year amongst people working at the time of diagnosis



What are the policy and system recommendations?

- 1. NHS Rare Diseases Advisory Group:** To support the implementation of the UK Rare Diseases Framework, specifically the commitment to ‘help patients get a diagnosis faster’:
 - a. Examine the opportunity for machine learning algorithms and artificial intelligence to detect PAH patients in primary care records and emergency care attendances.
- 2. NHS Specialised Respiratory Clinical Reference Group:** To guide patient-centred commissioning for PAH:
 - a. Invite a person with PAH to join the Clinical Reference Group as a Patient and Public Voice representative (current representatives cover cystic fibrosis, pulmonary fibrosis and brittle asthma, which have different needs and service structures).
 - b. Update the PH service specifications for specialist and shared care centres (last updated in 2013) to specify the components of comprehensive, ongoing patient-centred care. This should include psychological support, pulmonary rehabilitation, access to genetic services (including testing and counselling) for those with familial PAH, and links into local services for financial advice and community-based care (in line with the commitments in England’s 2024 Rare Diseases Action Plan).
 - c. Advocate for mention of PH/PAH in the *Adult breathlessness pathway* guidance published by NHS England in April 2023.
- 3. Integrated Care Boards:** To deliver a triple-win for conditions like PH, heart failure and respiratory disease, ensure local services are better set up to diagnose and manage breathlessness. To do this:
 - a. Maximise the opportunity of Community Diagnostic Centres (CDCs) by including vital services like phlebotomy, electrocardiography, echocardiography, CT scans and lung function testing.
 - b. Incorporate specialist oversight into local CDCs to support differentiation of the cause of breathlessness and timely referral onto an appropriate clinical pathway.
 - c. Structure services in line with the NHS *Adult breathlessness pathway* guidance.
- 4. NICE:** To support timely differentiation and diagnosis of PAH:
 - a. Update the clinical guidelines for more common conditions like heart failure, asthma and COPD to include reference to PAH in diagnostic algorithms as a potential cause for unexplained symptoms.
- 5. General Medical Council, General Pharmaceutical Council and Nursing and Midwifery Council:** To increase awareness amongst healthcare professionals, in line with the commitments in the UK Rare Diseases Framework:
 - a. Include content on PH (including PAH) in degrees and continued professional development modules for doctors, nurses, pharmacists and radiologists.



6. British Thoracic Society, British Cardiovascular Society, British Society for Rheumatology, British Society of Echocardiography, Royal College of General Practitioners and Royal College of Radiologists: To increase awareness of PAH in relevant specialties:

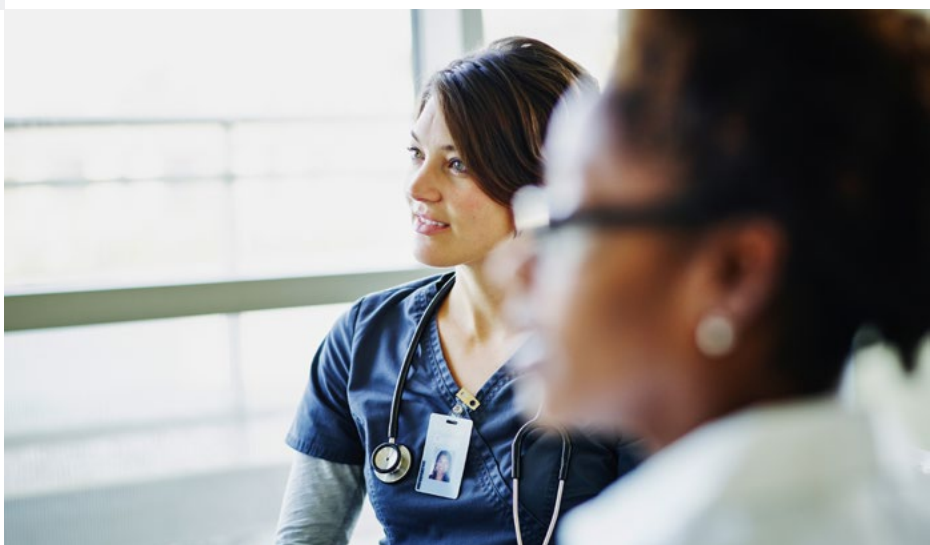
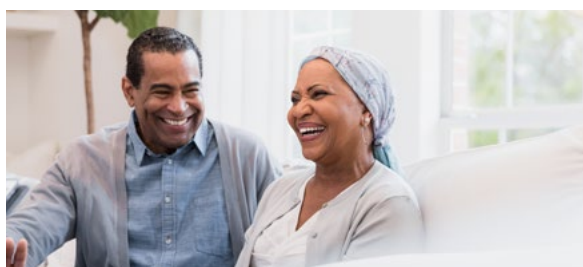
- a. Raise awareness of the NHS *Adult breathlessness pathway* guidance amongst your membership.
- b. Collaborate with the specialist PH centres to deliver continued professional development modules and events on PAH designed for general practitioners, radiologists, rheumatologists and the cardiovascular and respiratory workforce.
- c. Royal College of General Practitioners to develop a primary care screening tool for chronic unexplained breathlessness (e.g. when an inhaler does not improve symptoms).
- d. British Society of Echocardiography to incorporate content on PAH during echo study days for junior doctors.

7. Specialist Centres and Shared Care Centres: To lead the charge on realising this shared vision for PAH in the UK:

- a. Reinforcing established formal or informal links with local secondary hospitals to leverage your diagnostic expertise and support timely referral to specialist PH services. For example, this could include regular multidisciplinary meetings to discuss complex cases or assigning a designated point of contact in PH services for informal queries to guide referral.

8. Parliamentarians, especially those in cross-Party groups on respiratory or rare conditions: To amplify policy asks and personal stories of those living with PAH:

- a. Table parliamentary questions to the Secretary of State for Health and Social Care, or equivalent, asking:
 - What the Department is doing to support implementation of the NHS *Adult breathlessness pathway*.
 - What progress has been made to date on the implementation of the NHS *Adult breathlessness pathway*.
- b. Amplify the asks in this policy consensus statement with a letter to the Secretary of State for Health and Social Care, or equivalent.



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- **Mary Ferguson**, Communications Manager (PHA UK)
- **Professor David Kiely**, Consultant respiratory physician and Director of the Sheffield Pulmonary Vascular Disease Unit (Sheffield Teaching Hospitals)
- **Dr Colin Church**, Consultant respiratory physician and pulmonary vascular physician (Golden Jubilee Hospital)
- **Dr Neil Hamilton**, Consultant pharmacist in pulmonary vascular disease (Sheffield Teaching Hospitals)
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