

ORGANISATION OF PULMONARY HYPERTENSION (PH) DIAGNOSIS AND CARE

1 FIRST MEDICAL CONTACT: SUSPICION OF PH



PRIMARY CARE¹

- Medical history and comprehensive physical examination
- Screening tests (e.g. electrocardiogram (ECG), blood tests, chest X-ray)
 - Referral to a cardiac/respiratory specialist for further examination

Primary care physicians may misdiagnose people with PH with more common conditions, such as asthma or anxiety disorders.³

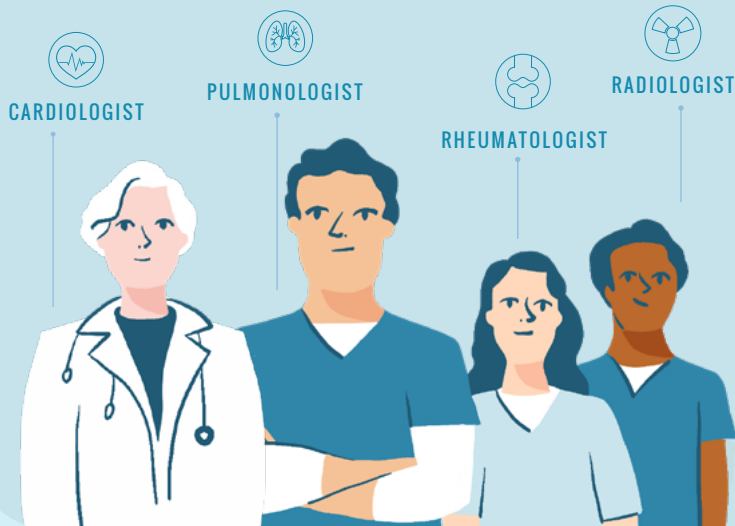


EMERGENCY DEPARTMENT²

- Screening tests (e.g. echocardiogram, blood tests, chest X-ray, computed tomography (CT) angiogram)
 - Referral to PH expert centre for diagnosis, follow-up management and/or critical care

People with PH visit the emergency department with severe symptoms, such as cardiac arrest, right heart failure, syncope, arrhythmia etc.²

2 SECONDARY CARE: SUSPICION OF PH



Key diagnostic test: echocardiography (to calculate PH probability)⁴

Other tests: ECG, chest X-ray, CT pulmonary angiography⁴



The specialist may not order the echocardiography or not consider the appropriate parameters to detect PH.⁵⁻⁷

The person with PH may instead be misdiagnosed with more common conditions, such as chronic obstructive pulmonary disease (COPD) or heart failure.^{3,5}

National comprehensive PH care

PH national referral expert centres are equipped with the workforce and services needed for an accurate diagnosis of PH and comprehensive treatment.⁸ Pulmonary arterial hypertension (PAH) is a rare and progressive – but treatable – form of PH.⁴ The European Society of Cardiology has established the quality indicators for PAH, including:⁸

A **multidisciplinary team** with expertise in PAH, including a nurse coordinator to provide person-centred care according to the latest guidelines and scientific advances.

Services including:

- specialist outpatient service
- intermediate/intensive care unit
- 24/7 emergency care
- interventional radiology unit
- cardiac anaesthesia and extracorporeal membrane oxygenation (ECMO)
- access to cardiothoracic and vascular surgery

Other responsibilities of PH national and shared care centres:⁸



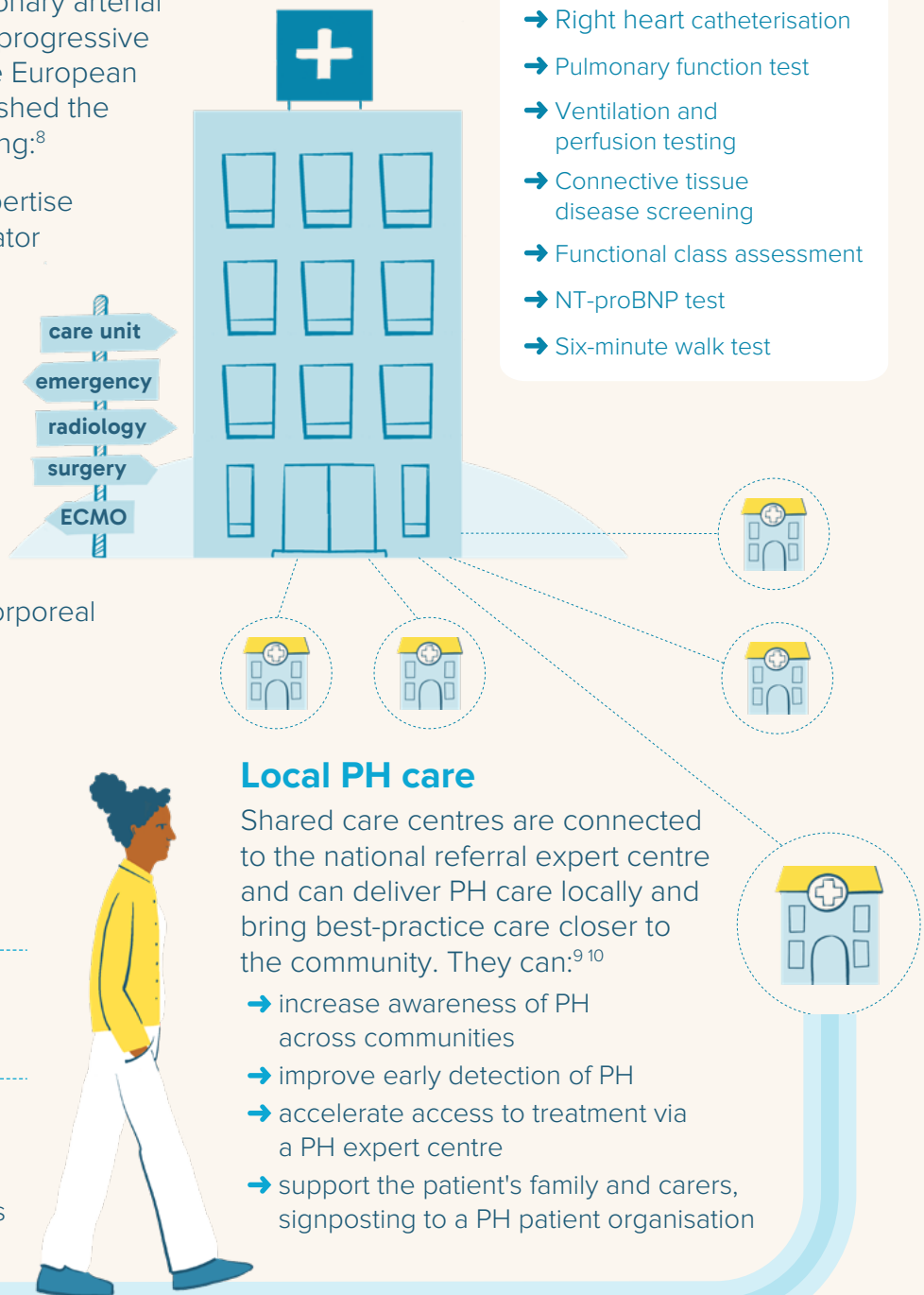
Participation in a national or international registry



Collection of data and undertaking of research



Education of, and shared decision-making between, patients and healthcare professionals around PH



Tests at the time of the diagnostic work-up and risk stratification for PAH:⁸

- Echocardiography
- CT angiogram
- Right heart catheterisation
- Pulmonary function test
- Ventilation and perfusion testing
- Connective tissue disease screening
- Functional class assessment
- NT-proBNP test
- Six-minute walk test

Local PH care

Shared care centres are connected to the national referral expert centre and can deliver PH care locally and bring best-practice care closer to the community. They can:^{9 10}

- increase awareness of PH across communities
- improve early detection of PH
- accelerate access to treatment via a PH expert centre
- support the patient's family and carers, signposting to a PH patient organisation

This document is not intended as educational material. All stakeholders should consult local formal guidelines, and their appropriate scientific and regulatory authorities, in any clinical decision or system design.

REFERENCES

1. Lua S, Church C. 2022. *Prescriber* 33(11-12): 11-18
2. Hofsfield R, Archer-Chicko C, Houston T, et al. 2018. *Adv Emerg Nurs J* 40(4): 246-59
3. Armstrong I, Rochnia N, Harries C, et al. 2012. *BMJ Open* 2(2): e000806
4. Humbert M, Kovacs G, Hoeper MM, et al. 2022. *Eur Heart J* 43(38): 3618-731
5. Small M, Perchenet L, Bennett A, et al. 2024. *Ther Adv Respir Dis* 18: 17534666231218886
6. Weatherald J, Humbert M. 2020. *Respirology* 25(8): 790-92
7. Kiely DG, Lawrie A, Humbert M. 2019. *Eur Heart J Suppl* 21(Suppl K): K9-k20
8. Aktaa S, Gale CP, Brida M, et al. 2023. *Eur J Heart Fail* 25(4): 469-77
9. Suntharalingam J. 2024. Interview [video call]. 25/01/24
10. Suntharalingam J, Ross RM, Easaw J, et al. 2016. *Clin Med (Lond)* 16(2): 135-41

The Health Policy Partnership

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