

**UNEXPLAINED  
BREATHLESSNESS?**



Check  
**RIGHT  
HEART**

**ACT RIGHT NOW**

**KEY FACTS**

Patients can wait **2.5 YEARS** on average for a PAH diagnosis from the onset of symptoms.<sup>3,4</sup>

By this time, **≥50 %** of pulmonary circulation may already be compromised.<sup>5</sup>

Symptoms can be **EASILY MISTAKEN** for other conditions such as asthma.<sup>1,4</sup>

**PH SYMPTOMS ARE NON-SPECIFIC AND INCLUDE BREATHLESSNESS, FATIGUE, ANGINA AND SYNCOPE.**<sup>1,6</sup>

Pulmonary hypertension (PH) is a haemodynamic and pathophysiological disorder found in multiple clinical conditions.<sup>1</sup> One type of PH – called pulmonary arterial hypertension (PAH) – is deadly if left untreated, and diagnosis is often delayed.<sup>2,3</sup>

**FIND YOUR NEAREST  
PH EXPERT CENTRE**

If you suspect PH, urgently refer your patient to a PH expert centre for further investigation.

**PULMONARY HYPERTENSION AND HEART LUNG TRANSPLANT UNIT**

**IV EPO**

St Vincent's Public  
and Private Hospital  
390-406 Victoria Street  
Darlinghurst NSW 2010

Professor Anne Keogh  
  
A/Professor Eugene Kotlyar

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**NEW SOUTH WALES PH EXPERT CENTRES**

**IV EPO**

Royal Prince Alfred  
Hospital

Level 11, KGV Building  
Missenden Road,  
Camperdown NSW 2050

T 02 9515 7410  
F 02 9550 6262



Westmead Hospital

Corner Hawkesbury Road and Darcy Road,  
Westmead NSW 2145

T 0457 305 841  
F 02 9986 1344



John Hunter Hospital

Lookout Road,  
New Lambton Heights NSW 2305

T 02 4985 5016  
F 02 4921 4210



Coffs Cardiology,  
Specialist Medical Centre

Level 2, Suite 208, 343 Pacific Highway,  
Coffs Harbour, NSW, 2450

T 02 6650 4100  
T 02 5626 8015  
F 02 6651 2606

Concord Repatriation  
General Hospital

Hospital Road,  
Concord NSW 2139

T 02 9767 5527  
F 02 9767 6994

Liverpool Hospital

75 Elizabeth Street,  
Liverpool NSW 2170

T 02 8738 4101  
F 02 8738 3054

Macquarie University  
Hospital

Suite 406, Level 4  
2 Technology Place,  
Macquarie University NSW 2109

T 02 9422 6002  
F 02 9475 1155

Nepean Hospital

Derby Street,  
Kingswood NSW 2747

T 02 4722 3001  
F 02 4722 5386

St Vincent's Clinic

Suite 710, 438 Victoria Street,  
Darlinghurst NSW 2010

T 02 8382 6150  
F 02 8382 6153

Sydney Adventist Hospital

185 Fox Valley Road,  
Wahroonga NSW 2076

T 02 9487 9111  
F 02 9480 9266

Wagga Wagga Rural  
Referral Hospital

260-280 Edward Street,  
Wagga Wagga NSW 2650

T 02 6932 1700  
F 02 6925 0699

**AUSTRALIAN CAPITAL TERRITORY PH EXPERT CENTRES**

**IV EPO**

Canberra Hospital

Yamba Drive,  
Garran ACT 2605

T 02 5124 2178  
T 02 5124 2371  
F 02 5124 2293

IV EPO, intravenous epoprostenol centre.

# IF YOU DETECT PH, IT COULD BE PAH

This guide can help you identify and refer patients at increased risk of PAH through three simple steps.

## STEP 1

IDENTIFY PATIENTS AT RISK

## STEP 2

DETECT PH

## STEP 3

CREATE A REFERRAL

### 1. IDENTIFY PATIENTS AT RISK

Early diagnosis of PAH is crucial, with intervention at this point likely to delay disease progression and improve patient outcomes.<sup>1,6</sup> Symptoms such as unexplained breathlessness require investigation.<sup>7</sup>

#### DO YOUR PATIENTS LIVE WITH ANY OF THESE SYMPTOMS?

- ✓ Unexplained dyspnoea
- ✚ PLUS any of the following
- ✓ Syncope
- ✓ Fatigue
- ✓ Weakness
- ✓ Chest pain
- ✓ Oedema
- ✓ Haemoptysis
- ✓ Abdominal distension
- ✓ Dry cough
- ✓ Exercise-induced nausea/vomiting

OR

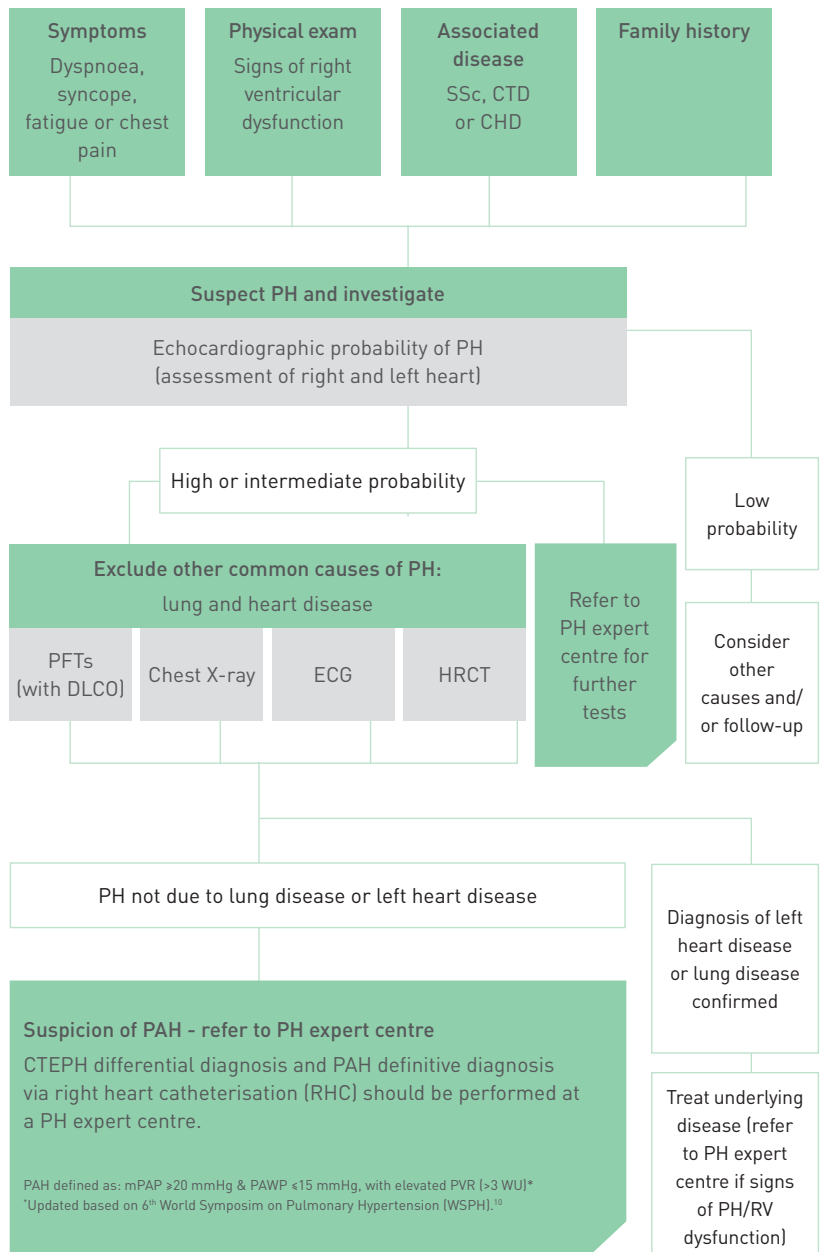
#### DO YOUR PATIENTS LIVE WITH ANY OF THESE CONDITIONS?

- ✓ CTD including SSc & SLE\*
- ✓ CHD
- ✓ HIV
- ✓ PoPH/Cirrhosis
- ✓ History of amphetamine use
- ✓ Family history of PAH

\*Patients with SSc should be screened annually for PAH.<sup>8</sup>

### INVESTIGATING PH IN PATIENTS AT RISK

Algorithm for assessment of suspected PH and referral to a PH expert centre<sup>1,9</sup>



Adapted from Galie N et al. 2016<sup>1</sup> and Frost A et al. 2019.<sup>9</sup>

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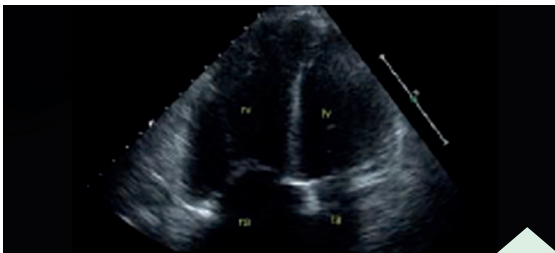
CREATE A REFERRAL

## 2. DETECT PH

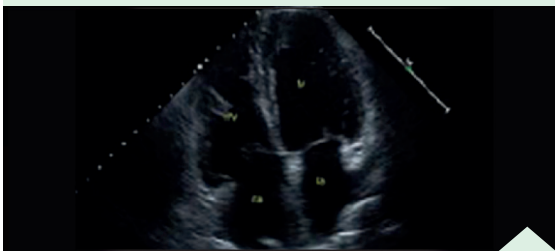
Echocardiography has become the first-line diagnostic and screening tool for PH.<sup>1,9,11</sup> **A comprehensive echocardiogram that looks at the right heart as well as the left should always be performed as part of a patient's work-up**, as it can provide evidence raising the suspicion of PH and build a case for definitive diagnosis at a PH expert centre that performs right heart catheterisation.<sup>1</sup>

**ECHOCARDIOGRAPHY (ECHO) SHOULD ALWAYS BE PERFORMED** when PH is suspected and may be used to infer a diagnosis.<sup>1</sup>

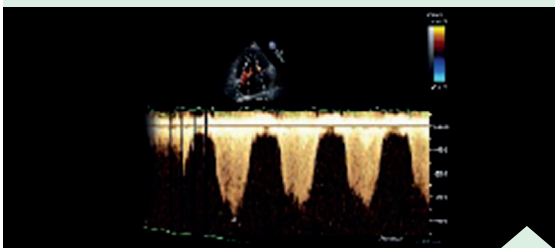
PH with evidence of cardiomegaly and enlarged pulmonary arteries:



Obvious PH by ECHO



Subtle PH by ECHO



Tricuspid regurgitant jet by stress ECHO

## NEXT STEPS WHEN ECHOCARDIOGRAM RAISES A HIGH OR INTERMEDIATE PROBABILITY OF PH

There are several other key investigations you can perform to raise suspicion of PH and help identify and classify its cause. Note that right heart catheterisation (RHC) is required to confirm a diagnosis of PAH, and should be performed by a PH expert centre.<sup>1</sup>



TO FIND OUT ABOUT FURTHER INVESTIGATIONS,

[VISIT SUSPECTPH.COM.AU](https://www.suspectph.com.au)

## CALCULATING RIGHT VENTRICULAR SYSTOLIC PRESSURE (RVSP)

RVSP is a derived parameter that is considered equal to systolic pulmonary artery pressure (sPAP) in most patients (in absence of pulmonary outflow obstruction).<sup>12</sup>

$$RVSP=4V^2 + RAP$$

V=velocity of tricuspid jet (m/s);  
RAP=right atrial pressure

*"When combined with the existing evidence, our findings now suggest that those presenting with estimated RVSPs >30.0 mmHg, no matter the eventual disease, should be carefully evaluated clinically, and where appropriate, further investigation should be undertaken to unmask disease that may benefit from disease-specific therapy."*

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### 3: CREATE A REFERRAL

Prompt diagnosis and treatment at a PH expert centre offers the best chance to delay disease progression and may improve patient outcomes for patients with PAH.<sup>1</sup>

**Right heart catheterisation (RHC)** is required for a definitive diagnosis of PAH. As RHC is technically demanding and can be associated with some serious complications, it should only be done at a PH expert centre.<sup>1</sup>

## IF YOU SUSPECT PH, URGENTLY REFER YOUR PATIENT TO A PH EXPERT CENTRE FOR FURTHER INVESTIGATION

FIND A PH EXPERT CENTRE AT [SUSPECTPH.COM.AU](https://suspectph.com.au)

### Abbreviations

**CHD:** congenital heart disease; **CTD:** connective tissue disease;  
**CTEPH:** chronic thromboembolic pulmonary hypertension;  
**DLCO:** diffusing capacity of the lung for carbon monoxide;  
**ECG:** electrocardiography; **ECHO:** echocardiogram; **HIV:** human immunodeficiency virus; **HRCT:** high-resolution computed tomography; **mPAP:** mean pulmonary artery pressure; **PAH:** pulmonary arterial hypertension; **PAWP:** pulmonary arterial wedge pressure; **PFTs:** pulmonary function tests; **PH:** pulmonary hypertension; **PoPH:** portopulmonary hypertension;  
**PVR:** pulmonary vascular resistance; **RAP:** right atrial pressure;  
**RHC:** right heart catheterisation; **RV:** right ventricle; **RVP:** right ventricular pressure; **RVSP:** right ventricular systolic pressure;  
**Ssc:** systemic sclerosis; **SLE:** systemic lupus erythematosus.

### References

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5. Lau EMT *et al.* *Eur Heart J* 2011;32:2489–98.
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7. Karnani NG *et al.* *Am Fam Physician* 2005;71:1529–37.
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