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How does PAH present in rheumatology?

Pulmonary arterial hypertension (PAH) is a well-known complication of connective tissue disease (CTD).¹ PAH associated with CTD (PAH-CTD) is most commonly seen in systemic sclerosis (SSc) and is a leading cause of death in these patients.² The [symptoms](#) and clinical presentation of PAH-CTD are very similar to idiopathic PAH.¹

Newly diagnosed patients with SSc should be screened for PAH at presentation and annually thereafter.¹

Due to the systemic nature of some CTDs, rheumatologists are also likely to encounter patients with other forms of pulmonary hypertension (PH), for example PH due to left heart or lung disease, which can complicate diagnosis and management especially if coexisting with PAH.³ Learn more about the different [types of PH](#).

What do rheumatologists need to look out for?

- In asymptomatic patients with SSc, annual screening for PAH is recommended using [resting echocardiography](#) combined with other measurements.¹ In certain patients with a disease duration >3 years, the DETECT algorithm is recommended.¹ Learn more about different [screening strategies](#)
- Symptoms that should prompt immediate consideration of PAH-CTD include unexplained dyspnoea, syncope, light-headedness, ankle oedema and chest pain^{1,2}
- Since the diagnosis of PAH-CTD can only be confirmed with right heart catheterisation (RHC), a thorough history, clinical examination and several non-invasive tests can be used to assess risk and need for RHC referral.¹
- Non-invasive tests that should be considered include:¹
 - Echocardiography (particularly if the tricuspid regurgitant velocity is >2.8 m/s or if there are features of right-sided chamber dilatation or dysfunction)²
 - Pulmonary function tests (particularly if the transfer factor of the lung for carbon dioxide [TLco] is falling, with a preserved forced vital capacity [FVC], leading to an FVC:TLco of >1.6)³
 - N-terminal pro-B-type natriuretic peptide (NT-proBNP). However, NT-proBNP is not specific to PAH-CTD and should not be used in isolation without other screening modalities³
 - Learn more about the different screening strategies and typical [investigation findings](#) in PAH associated with SSc (PAH-SSc)

Echocardiographic assessment of PAH

- Independent of underlying aetiology, signs of PH can be detected by echocardiography¹
- Resting echocardiography combined with other measurements is recommended to estimate the [echocardiographic probability of PH](#)¹
- Referral for RHC may be indicated to confirm the presence of PAH¹

DETECT*

DETECT is an evidence-based algorithm and is intended to help physicians in the UK screen patients with SSc with the goal to provide a recommendation for referral to echocardiography as a first step and, if applicable, for RHC as a second step for the diagnosis of PAH.

- The DETECT algorithm is recommended to identify asymptomatic patients with PAH (In adult patients with SSc of >3 years' duration, an FVC ≥40% and a DL_{CO} <60%)¹
- In patients with SSc, where breathlessness remains unexplained following non-invasive assessment, RHC is recommended to exclude PAH¹

You can access the DETECT algorithm by visiting the [detect website](#).

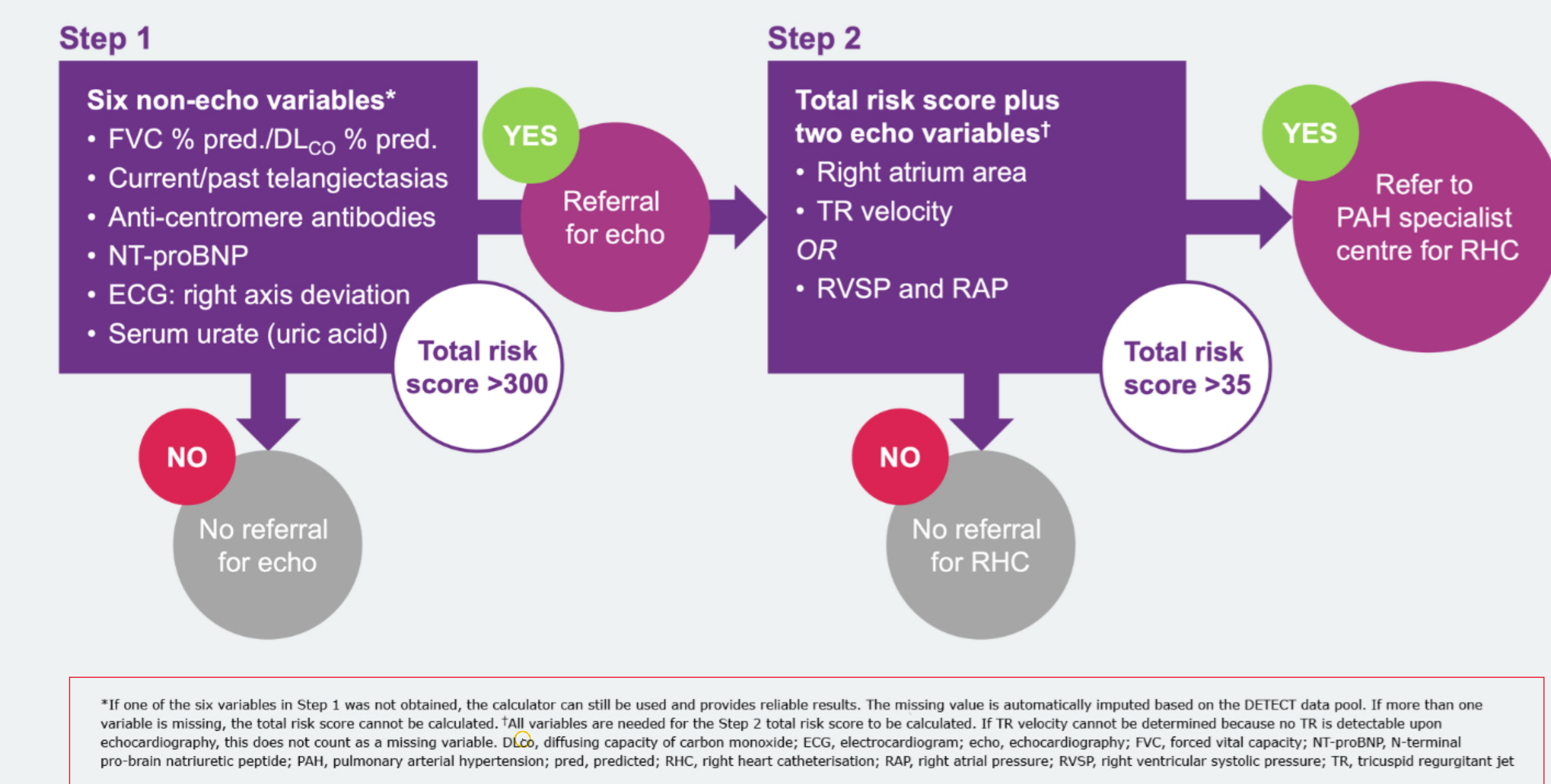


Figure 1. DETECT screening tool for PAH-SSc: an evidence-based algorithm to support you with your referral. Adapted from Coghlan JG, et al. Ann Rheum Dis. 2014; **23**:1340–9.⁴

DETECT disclaimers

*This PAH risk calculator is intended for use only by healthcare professionals in the UK. It has been developed on the basis of the DETECT study⁴ (this study was funded by Actelion Pharmaceuticals Ltd). Actelion was responsible for designing the study protocol, data collection and statistical analysis under the leadership of an independent study scientific committee. The DETECT PAH website is provided by Janssen Pharmaceutica N.V. The DETECT PAH website may not be used solely to make a diagnosis of PAH and does not replace specialist PAH clinical judgement. **Please read the Instructions for Use, warnings and precautions included within the website.**

Warnings and precautions: It is recommended to use the PAH risk calculator in centres specialised in the management of SSc and/or centres that work closely with these specialised SSc centres. The PAH risk calculator may not be used solely to make a diagnosis of PAH and does not replace specialist PAH clinical judgement. It should be used by specialists familiar with the disease, its diagnosis and treatment. It is not possible to determine the long-term performance of the algorithm nor to recommend how frequently patients should be screened, and the DETECT study included only patients with a DL_{CO} of <60%. Entering the wrong parameter values can lead to an incorrect risk score. To avoid misunderstanding of the parameters used for calculation, there is an information icon for each parameter in step 1 and step 2. Clicking on this icon will display the information for the corresponding parameter.

How do you refer a patient?

It is essential that this high-risk group is investigated and appropriately referred to one of the eight [adult specialist PH centres](#)⁸ in the UK whenever PAH-CTD is suspected.¹ The [DETECT algorithm](#) can help to indicate whether a patient requires RHC.⁴

The treatment of patients with PAH-CTD is complex;¹ however, early diagnosis and therapeutic intervention may offer an improved outlook for patients.^{5–7}

Key takeaways for rheumatologists

- PAH is a leading cause of death in patients with SSc²
- Patients with SSc should be screened annually for PAH, even if asymptomatic¹
- A combination of non-invasive tests may be performed to determine the need for RHC⁴

Related information

- [Detection of PAH in systemic sclerosis](#)
- [Practical guide to screening strategies](#)
- [SRUK annual testing booklet](#)

References

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