Pulmonary Hypertension Screening Adaptations for Adult Congenital Heart Disease Subtypes

Experts have devised PH screening adaptions for 4 subtypes of adult congenital heart disease. *Photo Credit: Gondelon/Science Source*

Despite the frequency with which pulmonary arterial hypertension (PAH) occurs in patients with adult congenital heart disease, there are no guidelines directing clinicians on echocardiographic screening for pulmonary hypertension (PH).1 In the absence of quality evidence, Dimopoulos and the CHAMPION Steering Committee (Congenital Heart Disease and Pulmonary Arterial Hypertension: Improving Outcomes Through Education and Research Networks) compiled evidence in a 25-study review and surveyed PH and congenital heart disease experts on the use of echocardiography for PH screening in patients with adult [**congenital heart disease**](https://www.pulmonologyadvisor.com/pulmonary-hypertension/pulmonary-arterial-hypertension-in-congenital-heart-disease/article/777463/).1

More than a decade ago pulmonologists recognized that early detection and treatment of PAH with a dual endothelin receptor antagonist in patients with congenital heart disease improved their exercise capacity and hemodynamic function.2 Even in patients with Eisenmenger syndrome — the most severe form of PAH — bosentan decreased the mean systemic arterial pressure, which may have slowed the progression to heart failure by reducing the strain on the right and left ventricles.2

**One Size Does Not Fit All**

Because of the heterogeneity of adult congenital heart disease, the CHAMPION Steering Committee acknowledged that standardized approaches for [**echocardiographic screening**](https://www.pulmonologyadvisor.com/pulmonary-hypertension/pulmonary-hypertension-predicted-in-interstitial-lung-disease/article/769973/) were impractical for 4 subgroups of congenital heart disease and offered screening adaptations1:

*Right ventricular outflow tract obstruction or pulmonary stenosis*

Because of anatomic abnormalities such as the unevenness of the tricuspid regurgitation (TR) gradient to the PA pressure, the ratio of the right ventricle/left ventricle diameters, and the right atrial size, the committee instead recommended that clinicians consider or evaluate the following1:

* The forward velocity/gradient across the pulmonary valve
* The peak Doppler velocity across the pulmonary-systemic vessels
* Significant pulmonary stenosis may influence the pulmonary regurgitation (PR) gradient

*Complex pulmonary atresia in tetralogy of Fallot*

In patients with this complex anatomy, Doppler with TR or mitral regurgitation (MR) cannot diagnose PH.1 Likewise, it is challenging to identify segmental PH with echocardiography. To screen for PH, the experts suggested using the following as potential PH markers:

* PA dilatation of hypertensive segments
* Estimated PA pressures with the Doppler interrogation of Blalock-Taussig shunts and collateral vessels*Unrepaired univentricular circulation*

Because neither TR nor MR Doppler can diagnose PH in patients with this type of congenital heart disease, the steering committee recommended the following screening adaptations:The PR gradient and gradient across systemic-to-pulmonary shunts indicate pressure gradient between the aorta and PA. PH may be present if the gradient is low

* If the PA is connected to a systemic ventricle, the low gradient across the pulmonary valve may indicate PH

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