

How to Evaluate Pericardial Disease in Congenital Heart Disease

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1. Congenital complete or partial absence of the pericardium

Complete or partial absence of the pericardium is rare. Although usually asymptomatic, patients can present with nonspecific symptoms such as dyspnea, light-headedness, and chest pain. Rarely, sudden death occurs due to herniation of the left atrium, left atrial appendage, or right atrium through the defect. Patients with complete absence of the pericardium usually are asymptomatic and require no treatment.

The diagnosis is not typically made by echocardiography, although unusual scanning windows, cardiac hypermobility, and abnormal ventricular motion may be present.

CT and MRI are diagnostic.

2. Pericardial Effusion in pulmonary arterial hypertension

Uncontrolled pulmonary arterial hypertension eventually leads to right ventricular failure with elevation of right atrial pressure and in some cases, the development of pericardial effusion. Postulated mechanism is venous/lymphatic obstruction with consequent cytokine release.

Echocardiography is also the gold standard for diagnosis of cardiac tamponade and the typical findings. Atypical signs of tamponade include isolated LA/left ventricular collapse and increased respiratory variation of transmitral inflow.

Cardiac MRI is also reliable and practical tool that can be used as part of the diagnostic work-up.

3. Pericardial and pleural effusion in congestive heart failure

Transudative pericardial effusion is not uncommon in patients with congestive heart failure. The prevalence of pericardial effusion has been reported at 12% to 20%. Pericardial effusion and/or pleural effusion can be expected when the right-sided filling pressure is elevated. In patients with biventricular failure, there is no evidence that elevated left-sided pressure, in the absence of elevated right-sided pressure, can cause a pericardial effusion.

4. Postpericardiotomy syndrome

Postpericardiotomy syndrome is a febrile illness secondary to an inflammatory reaction involving the pleura and pericardium. Pericardial effusions often accompany the syndrome and may develop into early or late postoperative cardiac tamponade and even recurrent cardiac tamponade. It is uncommon in patients younger than 2 years but the frequency increases in children and adults to as much as 30%.

5. Cardiac calcification in congenital heart disease

Heart calcification occurs in adult congenital heart disease patients, especially but not exclusively after intracardiac surgery. These calcifications can be associated with regional myocardial dysfunction, valve dysfunction, and sudden death.