Pericardial Agenesis with Resultant Pulmonary Hypertension

Travis R. Huffman, DO[1], E. Landon Smart, DO[2], Kris Pfirman, DO[1], Paden Vernon, MS-III[3], Aniruddha Singh, MD[4]

[1]University of Kentucky-Bowling Green, Cardiovascular Medicine Fellowship, Bowling Green, KY
[2]University of Kentucky-Bowling Green, Division of Internal Medicine, Bowling Green, KY
[3]University of Kentucky College of Medicine-Bowling Green Campus, Bowling Green, KY
[4]Med Center Health, Western Kentucky Heart and Lung Research Foundation and Education Trust, Bowling Green, KY

Introduction

The pericardium is a supportive layer that influences cardiac motion and orientation within the hemithorax. Absence of the pericardium, either complete or partial, is a rare congenital anomaly with an approximate incidence of less than 1/10,000, and is associated with other congenital defects in approximately 30-50% of cases. [1-2.]

While the complete absence of the pericardium is usually well tolerated and asymptomatic, partial absence can be both symptomatic and clinically significant. Symptoms of Congenitally Absent Pericardium (CAP), can include positional dyspnea and atypical chest pain. Clinically, patients can present with cardiac ischemia or sudden cardiac death, both of which are thought to be manifestations of cardiac herniation through an incomplete defect.[1,2,3,4.]

Case

An 80-year-old female with a history of diastolic heart failure, asthma and type II diabetes mellitus presented with encephalopathy and worsening dyspnea. She was subsequently found to be in septic shock secondary to a urinary tract infection, as well as in new onset atrial fibrillation.

HR: 76 **BP**: 87/55 **SpO2**: 97% on 2L NC

Admission ECG revealed new onset atrial fibrillation with controlled ventricular rate, and no evidence of ischemia.

2D Echocardiography, Figures 1 and 2, noted an atypical exam with apparent absence of the pericardial sac and significantly deviated viewing windows.

The parasternal long axis view was significant for a prominent right ventricle with marked right ventricular dilatation, and a posteriorly displaced left ventricle. The apical 4 chamber window was severely displaced both posteriorly and laterally, and was obtained posterior to the mid-axillary line.

2D Echo noted an ejection fraction of 50-55%, severe right ventricle dilation, moderate to severe tricuspid regurgitation, and estimated right ventricular systolic pressures of 40-50 mmHg, suggestive of moderate pulmonary hypertension.

Computed Topography of the Chest, Figures 3 and 4, showed bilateral pleural effusions with marked enlargement of the cardiac silhouette, aneurysmal appearance of the left ventricle, and severely enlarged main pulmonary arteries.

2D Echocardiogram

Figure 1. Parasternal Long Axis

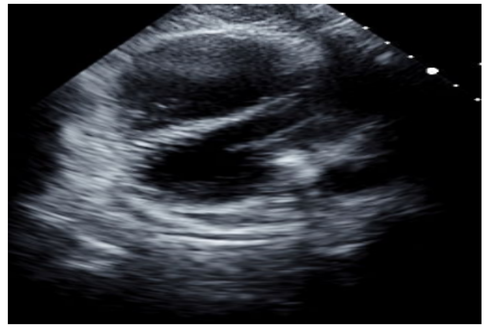


Figure 1. The parasternal long axis view reveals more of the right ventricle than normally visualized. The leftward shifting of the heart within the thoracic cavity may prompt concern for right ventricular enlargement as seen above. The right ventricle is still in normal orientation as located superior to the left ventricle and may be identified with the presence of the moderator band.

Figure 2: Apical 4-Chamber View

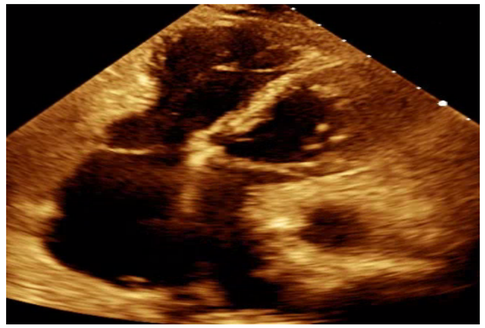


Figure 2. Paradoxical motion of the ventricular septum was present with what appeared to be an enlarged RV during systole, as well as bi-atrial elongation. The pericardial space is non-existent. The pulmonary pressure did appear to indicate moderate pulmonary hypertension in our case, with severe tricuspid regurgitation. The continuous and pulse wave dopplers of the LVOT and aorta indicated appropriate velocities and gradients, consistent with what would be expected of a normally configured heart.

Computed Topography Chest

Figure 3. CT Chest Axial

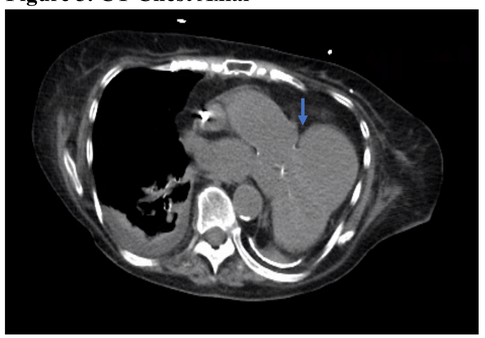
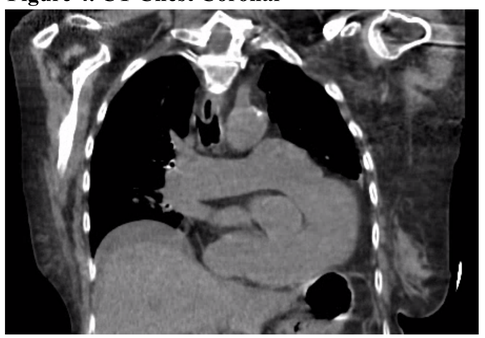


Figure 4. CT Chest Coronal



Figures 3 and 4. Lung tissue can be appreciated between the main pulmonary artery trunk and the aorta (blue arrow). The thin layer of pericardium appears to be absent in all views suggestive of pericardial agenesis. Levorotation of the heart is present and the heart appears to be excessively displaced leftward. The aorta in this case appears aneurysmal. The apex of the heart appears to be pointed posteriorly.

Discussion

Errors in the embryologic development of the pericardium can result in atrophy/agenesis of either the entire pericardium, or solely the right or left pericardial sac. The majority of patients with congenitally absent pericardium (CAP) are asymptomatic, therefore diagnosis is often made as an incidental finding.

Cardiac Computed Topography (CTT) and Cardiac Magnetic Resonance Imaging (CMR) provide better visualization of the thin pericardial membrane than surface echocardiography and can aid in diagnosis. Accurately identifying and describing the defect has both important diagnostic and prognostic implications, and will help guide long term management. CCT and CMR findings suggestive of pericardial agenesis include exaggerated levoposition of the heart, and interposition of lung tissue in the anterior space between aorta and pulmonary artery, or between the diaphragm and the base of the heart[5,6], as was present in our case.

Findings on 2D Echocardiography suggestive of pericardial agenesis include excessive cardiac motion in systole, described as “pendulum heart”, including paradoxical septal motion[5,6]. Also, the apical 4 chamber view can be significantly deviated both posteriorly and laterally, with elongated atria and widened ventricles creating the characteristic “tear-dropped” shape of the heart.[5,6,7]

In our case, the patient was found to have elevated right ventricle systolic pressure, as well as a severely enlarged main pulmonary artery suggestive of moderate pulmonary hypertension. While these findings only suggest the presence of pulmonary hypertension, it was suspected in our case that the significant deviation of the heart within the hemithorax resulted in compression atelectasis of the left lower lobe of the lung, physiologic pulmonary shunting, and increased pulmonary artery pressures. While enlargement of the main pulmonary arteries and tricuspid regurgitation are described in patients with congenitally absent pericardium, pulmonary hypertension is not explicitly described in the literature as a consequence of CAP.[6,7]

References

1. Yamano T, Sawada T, Sakamoto K, Nakamura T, Azuma A, Nakagawa M.. Magnetic resonance imaging differentiated partial from complete absence of the left pericardium in a case of leftward displacement of the heart. Circ J 2004;68:385–388.
2. Shah AB, Kronzon I. Congenital defects of the pericardium: a review. *Eur Heart J Cardiovasc Imaging*. 2015;16(8):821-8272014;22(1):36-39. doi:10.4250/jcu.2014.22.1.36. doi:10.1093/ehjci/jev119
3. Gatzoulis M.A., Munk M.D., Merchant N., Van Arsdell G.S., McCrindle B.W., and Webb G.D.: Isolated congenital absence of the pericardium: clinical presentation, diagnosis, and management. Ann Thorac Surg 2000; 69: pp. 1209-1215
4. Yared K, Baggish AL, Picard MH, Hoffmann U, Hung J. Multimodality imaging of pericardial diseases. *JACC Cardiovasc Imaging* 2010;3:650–60.
5. Klein A.L., Abbara S., Agler D.A., et al: American Society of Echocardiography clinical recommendations for multimodality cardiovascular imaging of patients with pericardial disease: endorsed by the Society for Cardiovascular Magnetic Resonance and Society of Cardiovascular Computed Tomography. J Am Soc Echocardiogr 2013; 26: 965–1012
6. David Lopez, Craig Asher. Progress in Cardiovascular Diseases, 2017-01-01, Volume 59, Issue 4, Pages 398-406.
7. Kim HJ, Cho YS, Cho GY, Choi SI. Congenital absence of the pericardium. *J Cardiovasc Ultrasound*.