CASE REPORT

Triphasic Mitral Inflow Pattern in a Patient with Congenital Absence of the Pericardium

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Abstract:
A healthy 19-year-old man was referred to our hospital due to an elongated left heart border on chest radiograph. Transthoracic echocardiography showed a posteriorly deviated left ventricular apex, increased right ventricular dimension, and hypokinesis of the interventricular septum. Pulsed Doppler echocardiogram revealed a triphasic mitral inflow pattern. Myocardial longitudinal strain rate imaging revealed that the early diastolic strain rate abruptly decreased to a negative value and then became positive at the left ventricular apex. In this case of an absent pericardium, the triphasic mitral inflow pattern might have been caused by an early diastolic shortening and subsequent elongation at the apex.

Key words: congenital absence of the pericardium, triphasic mitral inflow pattern, longitudinal strain rate


Introduction
Congenital absence of the pericardium is an exceedingly rare disorder in which the fibroserous membrane covering the heart is absent (1). Most patients with pericardial absence are symptom-free. We herein report a case of pericardial absence with a triphasic mitral inflow pattern caused by a particular left ventricular motion.

Case Report
A healthy 19-year-old male college student was referred to our hospital because of an abnormal chest radiographic finding. On a physical examination, the apical impulse was laterally displaced. The results of a blood sample analysis were normal. A 12-lead electrocardiogram showed a poor R wave progression (Fig. 1, left panel, arrows). A chest radiograph showed straightening and elongation of the left heart border and an obscured right heart border (Fig. 1, right panel). Computed tomographic scans of the chest suggested leftward deviation of the right atrium and posterior displacement of left ventricle (Fig. 2). The left pericardium seemed absent, as we noted no bright or linear structure of pericardium outside of the epicardial adipose tissue (arrows) (1). Two-dimensional transthoracic parasternal echocardiogram showed a posteriorly deviated left ventricular apex. The right ventricular dimension appeared enlarged, and the interventricular septum seemed hypokinetic (Fig. 3). A left ventricular M-mode echocardiogram showed an increased right ventricular dimension and hypokinesis of the interventricular septum (Fig. 3, L, upper panel). However, the interventricular septal wall motion seemed to increase from the left lateral decubitus position (Fig. 4, L, upper panel) to the supine position (Fig. 4, S, middle panel). Furthermore, the interventricular wall motion seemed normal, and the dimensions of the right and left ventricles decreased with the patient in the right lateral decubitus position (Fig. 4, R, lower panel). In this position, apical views were not obtained, probably due to the increased distance between the left chest wall and the heart.

Pulsed Doppler and pulsed tissue Doppler echocardiograms showed a triphasic mitral inflow pattern without a significant mid-diastolic wave at the septal annulus (Fig. 5). Myocardial longitudinal strain rate imaging from an apical four-chamber view showed that the early diastolic strain rate sharply decreased to a negative value and then increased to a positive value during mid-diastole at the left ventricular...
Discussion

Diagnosis of pericardial absence

In the present case, 12-lead electrocardiogram showed poor R wave progression and leftward displacement of the precordial transitional zone. Chest radiograph showed straightening and elongation of the left heart border (Snoopy sign) and loss of the right heart border (obscured by the spine) (Fig. 1, right panel) (1). A computed tomographic scan of the chest showed displacement of the heart posteriorly into the left hemithorax (Fig. 1, right, lower panel). Computed tomographic scans of the chest suggested leftward deviation of the right atrium and posterior displacement of the left ventricle. In addition, the left pericardium seemed absent, as we noted no bright or linear structure of pericardium outside of the epicardial adipose tissue. These findings were suggestive of an absent pericardium.

The transthoracic echocardiogram showed a posteriorly deviated left ventricular apex, an increased right ventricular dimension and hypokinesis of the interventricular septum. With the patient in the right lateral decubitus position, the right ventricular dimension and hypokinesis of the interventricular septum disappeared. Thus, the case was diagnosed as pericardial absence.

Congenital absence of the pericardium (1/10,000 autopsies) can present as left (70%), right (17%) or total bilateral (rare) absence (2). In the present case, chest radiograph
Figure 3. Parasternal long axis views in end-diastole (left panel) and end-systole (right panel) showing the posteriorly located left ventricular apex, enlarged right ventricle and paradoxical movement of the interventricular septum. Normal systolic thickening of the interventricular septum is present.

showed no significant interposition by the lung tissue between the aorta and main pulmonary artery. Therefore, this case might have been a case of partial left pericardial absence.

Mechanisms of the triphasic mitral inflow pattern

In the present case, pulsed Doppler echocardiogram showed a triphasic mitral inflow pattern. Although the details regarding the mechanism of this pattern remain unclear, a previous study described the hemodynamics of triphasic mitral inflow in left ventricular failure (3). In diastolic heart failure, low distensibility and high preload cause elevation of the left ventricular early diastolic pressure and reduce the mitral inflow. The left ventricular mid-diastolic pressure then decreases, and the mid-diastolic flow increases (Fig. 8, left panel).

In the present case, the longitudinal strain rate obtained from the apical four-chamber view revealed that the early diastolic strain rate was higher than in other segments and sharply decreased to a negative value before increasing to a positive value. One proposed mechanism for this is that apical active relaxation may be exaggerated without the restriction of the pericardium. The apex may then shrink to the length of other segments, thereby causing a decrease in the mitral inflow. The left ventricular mid-diastolic pressure may then decrease and the mid-diastolic flow increase (Fig. 8, right panel).

The findings in the present case also suggest that normal pericardium may equalize the left ventricular early diastole with uniform longitudinal strain.

In conclusion, we encountered a case of absent pericardium with a triphasic mitral inflow pattern that may have been caused by exaggerated active relaxation at the apex.
Figure 5. Pulsed Doppler echocardiogram at the mitral inflow (upper panel) and tissue Doppler echocardiogram at the mitral annulus (lower panel) with the patient in the left lateral decubitus position (L). Although the mitral inflow pattern is triphasic (orange arrows, upper panel), the mitral annular motion pattern is biphasic (white arrows, lower panel).

Figure 6. Myocardial longitudinal strain rate imaging from an apical four-chamber view in the left lateral decubitus position (L). The longitudinal strain rate obtained in this view revealed that the early diastolic strain rate was higher than in other segments and sharply decreased to a negative value (a) before increasing to a positive value (b).
Figure 7. Pulsed Doppler echocardiogram at the mitral inflow (upper panel) and tissue Doppler echocardiogram at the mitral annulus (lower panel) with the patient in the supine position (S). The mitral inflow pattern is also triphasic (orange arrows, upper panel). The mitral annular motion is biphasic.

Figure 8. Myocardial longitudinal strain rate imaging from an apical four-chamber view in the supine position (S). The early diastolic strain rate at the left ventricular apex decreases to a negative value during mid-diastole (a) and then increases to a positive value thereafter (b). This phenomenon seems subtle compared with observations in the left lateral decubitus position.
Figure 9. Possible mechanisms of the mid-diastolic flow in diastolic failure (left panel) and pericardial absence (right panel). In diastolic failure, low distensibility and high preload cause elevation of the left ventricular early diastolic pressure and decrease the mitral inflow. The left ventricular mid-diastolic pressure then decreases, and the mid-diastolic flow increases. In pericardial absence, apical active relaxation may be exaggerated without the restriction of the pericardium. The apex may then shrink to the length of other segments, causing a decrease in the mitral inflow. The left ventricular mid-diastolic pressure may then decrease and the mid-diastolic flow increase.

The authors state that they have no Conflict of Interest (COI).

References


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