

# CARDIOVASCULAR IMAGES

A joint publication of the Department of Radiology and Heart Center

## Congenital Absence of the Pericardium

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### Clinical History

A 57-year-old woman developed tachycardia during an elective hysterectomy and suffered from perioperative hypotension and mildly elevated cardiac biomarkers. Echocardiography was interpreted as severe inferior, septal, anterior, and apical hypokinesis. Subsequent cardiac catheterization revealed no obstructive coronary artery disease but raised the possibility of anomalous origin of the left main coronary artery from the right coronary cusp (Figure 1), and left ventriculography demonstrated basal hypokinesis. Her biomarkers returned to normal, and no action was taken. Several months later she was referred to cardiology clinic with vague complaints of atypical chest pain. Cardiac CT was requested to exclude a malignant interarterial course of the presumed anomalous left coronary artery.

### Findings

Cardiac CT angiography demonstrated no evidence of anomalous coronary arteries (Figure 2). Rather, the heart was found to be rotated leftwards on its vertical axis with the cardiac apex located posteriorly in the left hemithorax (Figure 3). The pericardium was not visualized. There were no regional or focal wall motion deficits, with preserved global systolic function. There was engorgement of the left lower lobe pulmonary veins. The findings were diagnostic of congenital absence of the pericardium.

### Discussion

Congenital absence of the pericardium is an uncommon anomaly, and can be either complete or partial (more commonly left-sided). The pathogenesis is believed to be secondary to an in utero vascular insult. Blood supply to the pericardium is via the musculophrenic branch of the internal mammary artery. The inferior pericardium is attached via the IVC; the superior pericardium is attached via the proximal pulmonary veins. Oftentimes patients are completely asymptomatic with the anomaly being discovered only incidentally, whereas symptomatic patients have been reported as experiencing intermittent stabbing chest pain.

Classic imaging findings include leftward deviation of the heart, posterior orientation of the cardiac apex and interposition of lung between the ascending aorta and the pulmonary artery in the expected region of the superior aortic pericardial recess (Figure 4).

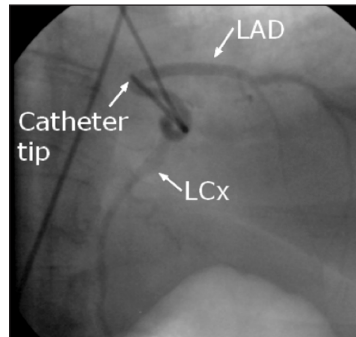


Figure 1

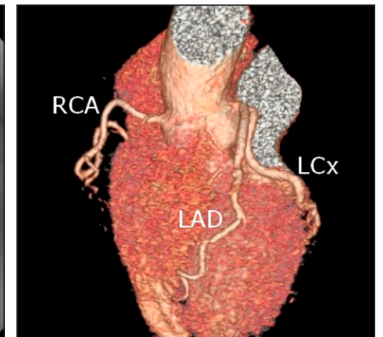


Figure 2

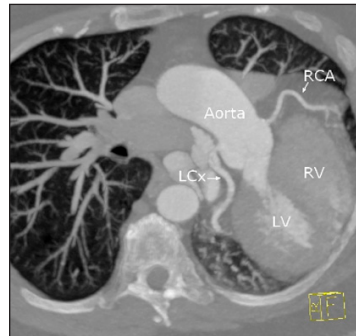


Figure 3

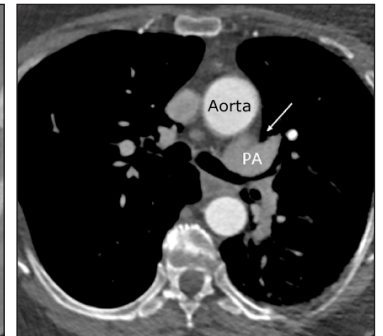


Figure 4

**Figure 1:** Apparent anomalous origin of the left main coronary artery from the right coronary cusp reported at the time of cardiac catheterization. The catheter tip is oriented towards the right, yet the left coronary system opacifies.

**Figure 2:** Although the heart is in an unusual orientation within the chest, there is no evidence of anomalous coronary arteries. The left main coronary artery arises from the left coronary cusp.

**Figure 3:** Leftward deviation of the heart with posterior orientation of the cardiac apex. The right ventricle therefore lies to the left and lateral to the left ventricle.

**Figure 4:** Interposition of lung between the ascending aorta and the pulmonary artery (PA) in the expected region of the superior aortic pericardial recess (arrow).

### REFERENCES

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