A 38-year-old woman with ill-defined chest pain had an abnormal x-ray 8 years before her only pregnancy. She had been unable to bear down during delivery, but gestation was otherwise uncomplicated, yielding a normal female offspring. She subsequently experienced left-sided chest pain that awakened her from sleep when turning into either a right or left lateral decubitus position. The pain was pulsating, stabbing, heavy, moderate in severity, and sometimes lasted for hours. Thoracic MRI in the supine and left lateral recumbent positions diagnosed congenital complete absence of the pericardium (Figure 1, left). The pain was ascribed to torsion at the thoracic inlet because of striking positional changes (mobility) of the heart (Figure 1, left). Congenital complete absence of the pericardium was confirmed during an operation designed to stabilize the cardiac position (Figure 1, right, and Figure 2). The heart was encapsulated in a Gore-Tex sack. The postoperative magnetic resonance images are shown in Figures 1 and 2. Figure 3 shows hypoplasia of the left lung and left pulmonary artery, which sometimes coexist with congenital complete absence of the pericardium.

Figure 1. Axial views. The preoperative (PreOp) image shows the mobile, unfixed heart. The postoperative (PostOp) image shows adhesion (fixation) of the lateral wall of the right ventricle to the chest wall (arrows) after surgical anchoring. RA indicates right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; and Ao, aorta.

Figure 2. Dynamic images obtained in a horizontal long axis (4-chamber) view in diastole and systole with the patient positioned on her right side. These images illustrate the absence of significant cardiac motion after surgical fixation of the heart to the chest wall (arrows). Abbreviations as in Figure 1.
Figure 3. Three-dimensional reconstruction of gadolinium-enhanced MR angiography of the heart and great arteries in the anterior (ANT) and left anterior oblique (LAO) views. The left anterior oblique view shows the hypoplastic left pulmonary artery (LPA). RPA indicates right pulmonary artery; Ao, aorta; PA, pulmonary artery; S, superior; and I, inferior.
Congenital Complete Absence of the Pericardium
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