Ectopia cordis is a rare congenital lesion affecting between 5.5 and 7.9 children per one million live births. Depending on the location of the heart, the lesion can be classified as cervical, thoracic, thoracoabdominal, or abdominal. Ectopia cordis is often associated with other midline defects, including the constellation of defects know as the pentalogy of Cantrell. The components of pentalogy of Cantrell are ectopia cordis, sternal cleft, midline abdominal defect or omphalocele, a pericardial defect allowing communication between the pericardial and peritoneal cavities, and one or more cardiac defects.

Management of ectopia cordis entails coverage of the bare heart and palliation or correction of any associated heart defects. Placement of the heart into the thorax with sternal or thoracic reconstruction is desirable when possible. Although previous reports have shown very poor outcomes for infants with ectopia cordis and associated cardiac defects, more recent reports have demonstrated improved survivals. Whereas this patient’s chest radiograph appeared normal (Figure 1), physical exam, Doppler/echocardiography, and cardiac catheterization revealed ectopia cordis, right atrioventricular valve atresia, and a single morphologically left ventricle (LV) entirely below the level of the diaphragm (Figures 2 and 3). She subsequently underwent successful closure of her pericardium and abdominal fascia with polytetrafluoroethylene patches and primary skin closure. Currently, her pulmonary and systemic circulations are well balanced, with oxygen saturations in the range of 85%. Her anticipated management will include a hemi-Fontan procedure at 6 months of age and ultimately a Fontan procedure at 18 to 24 months.

Figure 1. A chest roentgenogram with normal appearing cardiac silhouette, apparent levocardia, and normal pulmonary vascular markings.

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Circulation encourages readers to submit cardiovascular images to the Circulation Editorial Office, St Luke’s Episcopal Hospital/Texas Heart Institute, 6720 Bertner Ave, MCI-267, Houston, TX 77030.

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Figure 2. Doppler/echocardiography and cardiac catheterization revealed right atrioventricular valve (tricuspid) atresia with a single morphologically LV entirely below the level of the diaphragm. Only the atria, great vessels, and left atrioventricular valve were within the thorax. This LV angiogram demonstrates the single LV below the diaphragm. The aorta arises from the LV, whereas the small pulmonary artery arises from an outlet chamber fed by a bulboventricular foramen.

Figure 3. A 36-week gestation neonate delivered by caesarian section for a fetal diagnosis of pentalogy of Cantrell.
Normal Chest X-Ray
Richard G. Ohye and Thomas A. Kulik

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