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Ectopia cordis with right and left ventricular diverticula

A newborn girl who was delivered vaginally at 37 weeks of gestation exhibited double-outlet right ventricle, pulmonary stenosis and persistent left superior vena cava accompanied with ectopia cordis (online video 1: shows her heart beating outside the body and the anterior interventricular sulcus). The case fulfilled the diagnostic criteria of Cantrell’s pentalogy. Half of each of the right and left cardiac ventricles was exposed. An emergency operation was performed to place the exposed portion of the heart into the internal thoracic cavity. Expanded polytetrafluoroethylene (e-PTFE) was used as a substitute for the pericardium. Subsequently, skin flaps were designed on both the sides and sutured to each other such that they covered the e-PTFE patch-layered heart. After 2 months, modified Blalock-Taussig shunt was created on the right side to treat cyanosis. Computed tomography of the chest, which was performed after the palliative operation, showed the rightward-pointing heart apex and the presence of both right and left ventricular diverticula (panel A–B).

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Additional video published online only. To view it, please visit the journal online (http://heart.bmj.com).

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Panel A. The frontal view of the three-dimensional computed tomography shows sternum dysplasia and the rightward-pointing heart apex with the presence of both right (Rt.) and left ventricular diverticula. B. The three-dimensional computed tomography focusing on cardiac anatomy shows bilateral superior vena cava (SVC) and double outlet right ventricle with subpulmonary stenosis. A left anterior descending coronary artery passes in the middle of the two ventricular diverticula.