Multidetector Computed Tomographic Angiography Imaging of Pentalogy of Cantrell

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A 4-day-old female mildly cyanotic neonate presented clinically with ventral herniation of the heart to the anterior abdominal wall and umbilical defect containing bowel loop (Figure [A] and Movie I in the online-only Data Supplement), raising suspicion of pentalogy of Cantrell. Echocardiogram of the ectopic heart, although technically difficult, revealed overlap of the ascending aorta with small-caliber partially visualized main pulmonary artery arising from the ventricle (Figure [B]). Multidetector computed tomographic angiography (MDCTA) confirmed the diagnosis of pentalogy of Cantrell, which consisted of ectopia cordis with rightward apex containing ventrally herniated single ventricle with right-sided atrioventricular valve atresia protruding through the defective anterior diaphragm anterior to the liver, omphalocele, lower sternal and ventral thoracoabdominal wall defects, and multiple cardiac anomalies (Figure [C through E] and Movie II in the online-only Data Supplement). Cardiovascular anomalies included single ventricle with right-sided atrioventricular valve atresia giving rise to tetralogy of Fallot–type conotruncus with diffusely narrowed main pulmonary artery. Additional anomalies included large secundum atrial septal defect, absent left pulmonary artery originating from the undersurface of the aortic arch by way of patent ductus arteriosus, and left aortic arch with aberrant right subclavian artery (Figure [C through E] and Movie II in the online-only Data Supplement). No abnormal systemic venous drainage or aortic coarctation was found. The complex combination of the aforementioned cardiovascular anomalies suggested a poor prognosis. Because pentalogy of Cantrell can be fatal without surgery, surgery was decided on after discussion with the patient’s parents. At the initial stage, surgeons attempted to cover the unprotected heart with skin with the understanding of a requirement for repair of intracardiac defects at a later age. However, while the heart was shifted caudally during the initial stage operation, the patient experienced progressive hemodynamic instability, which unfortunately resulted in the patient’s death. We believe that the unfortunate outcome may be secondary to unexpected great vessel compression or kinking during the surgical procedure.

Discussion

The first description of the pentalogy of Cantrell was given by Cantrell et al.1 The pentalogy of Cantrell is a rare fatal anomaly with prevalence ranging between 1:65,000 and 1:200,000 births and can manifest with variable degrees of severity, with the classic complete type comprising 5 components: lower sternum defect, omphalocele, anterior diaphragmatic defect, diaphragmatic pericardial defect, and variable congenital cardiac anomalies.1,2 Most cases are sporadic with unknown pathogenesis. The possible underlying pathogenesis is developmental failure of the lateral mesoderm segment during 14 to 18 days of gestational age.2

The expression of the pentalogy of Cantrell varies in severity from complete to incomplete syndrome; however, anterior sternal defect is always present.3 Ventricular septal defect is the most common cardiac-associated anomaly; other associated congenital cardiac lesions include atrial septal defect, pulmonary valve stenosis, tetralogy of Fallot, dextrocardia, anomalous pulmonary venous connection, tricuspid atresia, and truncus arteriosus.3 Our case represented the full spectrum of the pentalogy of Cantrell. Our case presented with a complex combination of thoracoabdominal ectopia cordis in association with single ventricle, atrial septal defect, and absent left pulmonary artery originating from the undersurface of the aortic arch by way of patent ductus arteriosus; thus, the prognosis was poor.

The pentalogy of Cantrell can be diagnosed earliest during a first-trimester antenatal sonogram.2 However, because of lack of awareness, our patient had never undergone an antenatal sonogram. The precise anatomic identification of the components of the pentalogy of Cantrell can be difficult because of the spatial complexity of the anomaly, evaluation of which is...
extremely important for treatment planning and prognostication. Multimodality imaging with echocardiogram, magnetic resonance imaging, and MDCTA remains helpful for pretreatment planning. MDCTA was chosen for its capacity to demonstrate the segmental anatomy and the position of the heart relative to the bones of the thoracic cage.

Because the prognosis depends on the severity of the pentalogy of Cantrell, precise anatomic description is mandatory; however, the patient with pentalogy of Cantrell will eventually die without surgery. Management is surgical, consisting of palliative repair of the ventral hernia and diaphragmatic defect and palliative or corrective repair of the cardiovascular anomalies. However, mortality is still high for the complex surgeries done for highly severe cases.

In summary, we have demonstrated a complete case of pentalogy of Cantrell with clinical and state-of-the-art MDCTA imaging. MDCTA with 3-dimensional volume rendering and maximum-intensity projection postprocessing techniques can be highly useful in accurate pretreatment assessment of the complex anomalies associated with pentalogy of Cantrell.

Disclosures
None.

References
Figure. A, Clinical picture of the patient reveals large ventral herniation of the heart into lower chest and upper abdomen (arrow) along with ventral bowel herniation through the umbilicus (arrowhead) suggestive of omphalocele. B, Echocardiogram through the ectopia cordis demonstrates overlap of ascending aorta (aAO) with small-caliber partially visualized main pulmonary artery (mPA) arising from the ventricle. C, Sagittal 3-dimensional volume-rendered computed tomographic image reveals lower sternum defect (arrowhead), ventral ectopia cordis (bold arrow), and an omphalocele (thin arrow). D and E, Maximum-intensity projection (D) and volume-rendered images (E) demonstrate ventrally herniated single ventricle (SV) with rightward apex, giving rise to tetralogy of Fallot–type conotruncus and diffusely hypoplastic main pulmonary artery, which branches into the hypoplastic right pulmonary artery (triangular arrowhead) with absent left pulmonary artery originating from undersurface of aortic arch (thin arrow) by way of patent ductus arteriosus (bold arrow). Note patent ductus arteriosus (bold arrow) extending between the inferior aspect of aortic arch and the left pulmonary artery (thin arrow). Additional findings include aberrant right subclavian artery (short arrowhead) from the proximal descending aorta.
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**Movie Legend**

**Movie 1.** Clinical presentation of the patient’s chest and upper abdomen. The recording reveals large ventral herniation of the heart to lower chest and upper abdomen along with omphalcele. Best viewed with Windows Media Player.

**Movie 2.** Computed tomography (CT) of the chest and upper abdomen demonstrating several cardiovascular abnormalities associated with Pentalogy of Cantrell. Movie clip of the axial CT images from the chest to the upper abdomen reveals several cardiovascular abnormalities associated with Pentalogy of Cantrell, including ectopia cordis, multiple cardiovascular anomalies along with defects in the lower sternum and ventral thoraco-abdominal walls. Cardiac defects include large secundum type anterior septal defect, a ventrally herniated single ventricle with rightward apex and right-sided atrioventricular valve atresia, giving rise to TOF-type conotruncus with diffusely narrowed main pulmonary artery. Several other cardiovascular anomalies are also noted, including absent left pulmonary artery originating from undersurface of aortic arch by way of patent ductus arteriosus and left aortic arch with aberrant right subclavian artery. Best viewed with Windows Media Player.