The Heart Surgery Forum #2006-1135
10 (6), 2007 [Epub October 2007]
doi: 10.1532/HSF98.20061135

Online address: http://cardenjennings.metapress.com

Multidetector Computed Tomography Findings of Ectopia Cordis and Other Components of Pentalogy of Cantrell: A Case Report

Mecit Kantarci,[1] Fadime Fil,[1] Naci Ceviz[2]

[1]Department of Radiology and [2]Division of Pediatric Cardiology, Medical Faculty, Atatürk University, Erzurum, Turkey

ABSTRACT

We report a case of ectopia cordis assessed with multidetector computed tomography (MDCT). Accurate and early diagnosis in components of pentalogy of Cantrell is vital for surgical planning and assessment of prognostic factors. Postnatal MDCT imaging allows for optimal assessment of the fetus with ectopia cordis, which has significant implications in terms of preoperative planning and obtaining prognostic information.

INTRODUCTION

Pentalogy of Cantrell is a rare thoracoabdominal disruption in the abdominal wall, diaphragm, pericardium, and sternum. It is a heart defect that was initially described as a specific entity by Cantrell and colleagues in 1958 [Cantrell 1958]. The estimated prevalence of pentalogy of Cantrell varies from 1 in 65,000 to 1 in 200,000 births, and it is currently classified as an anterior body wall midline developmental anomaly [Carmi 1992]. Most often, the cardiac lesion is a ventricular septal defect. In severe cases, the heart herniates through the diaphragmatic defect, causing thoracoabdominal ectopia cordis. Other associated congenital cardiac lesions may include an atrial septal defect, pulmonary valve stenosis, tetralogy of Fallot, dextrocardia, anomalous pulmonary venous connection, tricuspid atresia, and truncus arteriosus. An associated left ventricular apical aneurysm has been reported in several cases [Carmi 1992; Vasquez-Jimenez]. This report represents the importance of multidetector computed tomography (MDCT) in the assessment of ectopia cordis, other components of Cantrell pentalogy in preoperative planning, and in obtaining prognostic information.

CASE REPORT

A 20-year-old mother's first child, a female newborn, was delivered in the the 37th week of gestation by cesarean section, following the detection of an anomalous heartbeat. The birth weight was 3360 g. The mother denied any known exposure to teratogenic agents, and there was no other family history of birth defects. During her pregnancy, the mother had not undergone any imaging method to detect prenatal anomalies. After the birth, the newborn was referred to our center because of an ectopia cordis finding, and at that time a preoperative multidetector computed tomography (MDCT) was planned to evaluate the diaphragmatic defect. MDCT was performed using 16-detector row CT scanner (Aquillon; Toshiba Medical Systems, Tokyo, Japan).

Axial-sagittal maximum intensity projection (MIP) images and three-dimensional volume-rendering images were obtained. Sagittal MIP image (Figure 1) showed a thoracoabdominal wall defect, with the right and left ventricle herniating anteriorly, below a hipoplastic sternum, superior to the liver. There was a partial defect in the lower sternum and a large ventricular septal defect. The aorta overrode the ventricular

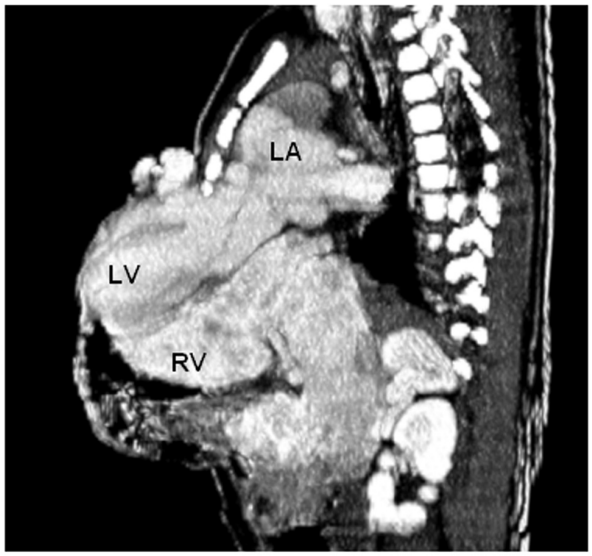


Figure 1. Sagittal maximal intensity projection image shows the right and left ventricle extending ventrally below a hypoplastic sternum. Presence of superior and inferior vena cava and the intrathoracic location of both atriums are also seen. RV indicates right ventricle. LV, left ventricle; LA, left atrium.

Received June 30, 2007; received in revised form July 19, 2007; accepted July 23, 2007.

Correspondence: Mecit Kantarci, MD, 200 Evler Mah, 14, Sok No 5, Dadaskent, Erzurum, Turkey; 90-442-3273802 (home); 90-442-2361212-1521 (work); fax: 90-442-2361301 (e-mail: akkanrad@hotmail.com).

septal defect, and the aortic root was dilated. Axial MIP image (Figure 2) showed the herniating ventricles running through the extrathoracic region. Three-dimensional volume rendering images (Figure 3) also showed the anterior thoracic cardiac displacement.

DISCUSSION

Cantrell's pentalogy, first described in 1958, consists of a lower sternal defect, anterior diaphragmatic defect, parietal pericardial defect, omphalocele, and herniation of the heart. Ectopia cordis, described as the herniation or extrusion of the heart, may occur either in the thorax or in the thoracoabdominal region [Martin 1992]. The hypothesis underlying this condition is abnormal development of the mesoderm at a very early stage in embryonic life, occurring prior to or immediately after differentiation of the primitive intraembryonic mesoderm into its splanchnic and somatic layers, because derivatives of both these layers are involved [Hornberger 1996; Di Bernardo 2004]. To date, no etiologic factors have been decribed. In approximately the 10th week of embryological life, the midgut, which had previously been extraabdominal, returns to the abdominal cavity and undergoes rotation, followed by fixation. The final structure of the abdominal wall is composed of 5 distinct body folds that fuse at the base of the umbilical cord. Failure of growth and fusion of the cephalic fold results in either isolated ectopia cordis or the pentalogy of Cantrell or one of its variants. The prognosis depends on significant extracardiac defects, complex cardiac anomalies, pulmonary hypoplasia, large abdominal wall defects, cerebral anomalies, and herniation of the bowel into the thoracic cavity, because all these would likely worsen the overall prognosis of these patients [Halbertsma 2002; Morales 2000]. Prenatal monitoring and diagnosis of ectopia cordis in utero are possible through obstetric ultrasonography and, once identified, ectopia cordis should prompt a search for other anomalies [Ghidini 1998]. Postnatal computed tomography has proven useful to estimate the thoracic volume in considering surgical intervention of the ectopic heart [Haynor 1984]. MDCT allows optimal assessment of the fetus with ectopia cordis and pentalogy of

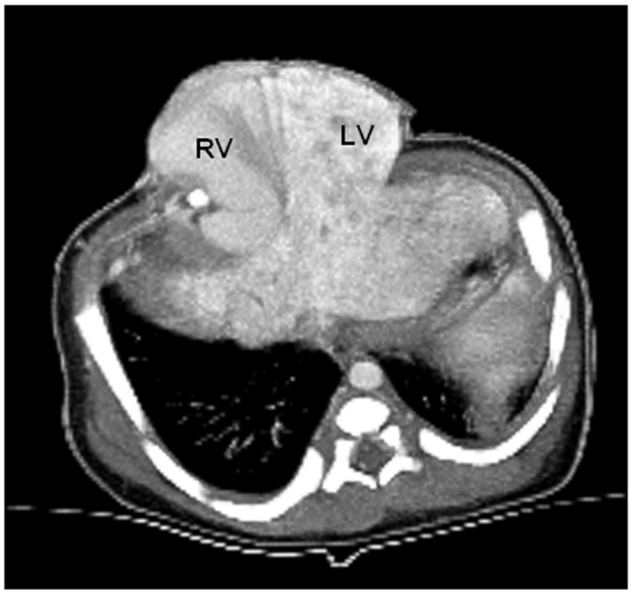
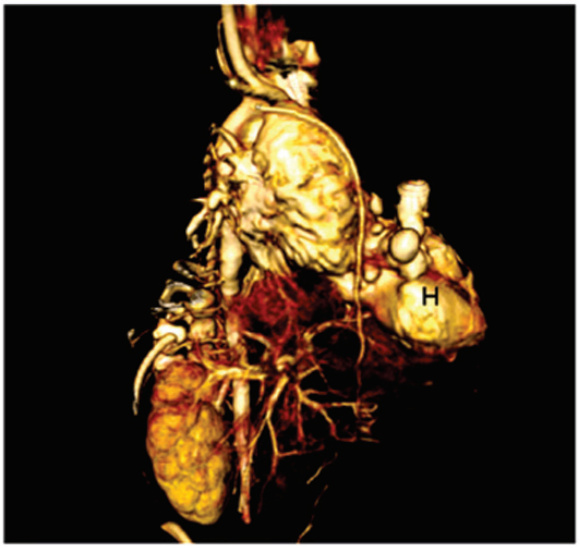
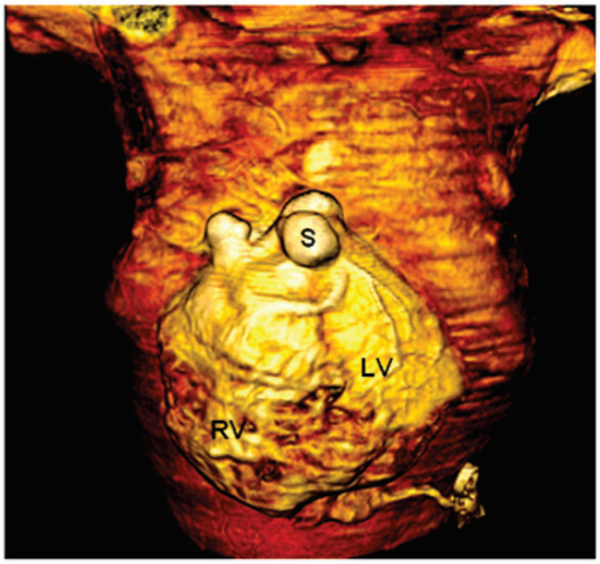


Figure 2. Axial maximal intensity projection image at the level of the midthorax demonstrating the ectopia cordis. RV indicates right ventricle; LV, left ventricle.



A



B

Figure 3. Sagittal (A) and coronal (B) three-dimensional volume rendering images show the anterior thoracic cardiac displacement (ectopia cordis). RV indicates right ventricle, LV, left ventricle; S, sternum; H, heart.

Cantrell. This has significant implications in terms of preoperative planning and obtaining prognostic information, particularly with the emergence of fetal surgical programs.

REFERENCES

Cantrell JR, Haller JA, Ravitch MM. 1958. A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium and heart. Surg Gynecol Obstet 107:602-14.

Carmi R, Boughman JA. 1992. Pentalogy of Cantrell and associated midline anomalies: a possible ventral midline developmental field. Am J Med Genet 42:90-5.

Di Bernardo S, Sekarski N, Meijboom E. 2004. Left ventricular diverticulum in a neonate with Cantrell syndrome. Heart 90:1320.

Ghidini A, Sirtori M, Romero R, Hobbins JC. 1998. Prenatal diagnosis of pentalogy of Cantrell. J Ultrasound Med 7:567-72.

Halbertsma FJ, van Oort A, van der Staak F. 2002. Cardiac diverticulum and omphalocele: Cantrell's pentalogy or syndrome. Cardiol Young 12:71-4.

Haynor DR, Shuman WP, Brewer DK, Mack LA. 1984. Imaging of fetal ectopia cordis: roles of sonography and computerized tomography. J Ultrasound Med 3:25-7.

Hornberger LK, Colan SD, Lock JE, Wessel DC, Mayer J. et al. 1996. Outcome of patients with ectopia cordis and significant intra-cardiac defects. Circulation 94(9 Suppl)II32-7.

Martin RA, Cunniff C, Erickson L, Jones KL. 1992. Pentalogy of Cantrell and ectopia cordis, a familial developmental field complex. Am J Med Genet 42:839-41.

Morales JM, Patel SG, Duff JA, Villareal RL, Simpson JW. 2000. Ectopia cordis and other midline defects. Ann Thorac Surg 70:111-114.

Vazquez-Jimenez JF, Eberhard GM, Muehler EG, Daebritz S, et al. 1998. Cantrell's syndrome: a challenge to the surgeon. Ann Thorac Surg 65:1178-85.