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Title: 64-slice MDCT Imaging of Endocardial Cushion Defect Associated with Other Cardiac and Extracardiac Abnormalities

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Key words: endocardial cushion defect, multi detector computed tomography, volume rendering view, congenital heart disease, heterotaxy syndrome

A 30-year-old man presented with heart failure and had very low heart rate (40~45). He had been diagnosed with a complete endocardial cushion defect (ECD) when he was 3 years old. He was referred for an indication of a permanent pace maker. Contrast-enhanced ECG-gated 64-slice MDCT was held to evaluate cardiac anatomy beforehand. Multi-planar reconstruction (MPR) image showed the common atrium without atrial septum, large ventricular septal defect and small right ventricle due to complete atrioventricular canal defect (Figure 1a and 1b). 3D-CT volume rendering imaging also revealed the patent ductus arteriosus (PDA), dilatation of the ascending aorta (Figure 2a), and the anomalous interarterial course of the right coronary artery running between the ascending aorta and pulmonary artery (Figure 2b), which were not noticed beforehand. This patient also had heterotaxy syndrome with polysplenia and azygos continuation. (Figure 3a and 3b)

ECD is the abnormal or inadequate fusion of the superior and inferior endocardial cushion, which normally occurs during the 5th week of gestation. ECD has broad spectrum of defects characterized by involvement of the atrial septum, ventricular septum and atrioventricular valves.

(1) MDCT is a good non-invasive imaging modality for the evaluation of the whole heart and great vessels (aorta and pulmonary artery, and PDA) simultaneously, with wide coverage. (2) MPR images enable accurate estimation of the size and location of a defect and evaluation of associated anomalies.

References

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Figure Legends:

Figure 1: Multiplanar reconstruction (MPR) images

Figure 1a (left): MPR image with both ventricles shows the common atrium (white arrow) without atrial septum and small right ventricle.

Supplement file; movie clip of Figure 1a (continuous 16 MPR images)

Figure 1b (right): Short axis MPR image shows straddling atrioventricular valve leaflets (black arrow) over ventricular septum with large ventricular septal defect (asterisk).

RV; right ventricle, LV; left ventricle

Figure 2: 3D-CT Volume Rendered (VR) images

3D-CT image (Figure 2a, left: left anterior view) shows the patent ductus arteriosus (black arrow) and dilated ascending aorta (asterisk), and another image without pulmonary artery (Figure 2b, right) shows the anomalous origin right coronary artery (black arrow).

Figure 3: Axial contrast-enhanced images obtained with cardiac CT

Figure 3a (left): Axial CT shows heterotaxy syndrome with polysplenia (asterisks). Dilated azygos vein (black arrow) runs beside the descending aorta (white arrow).

Figure 3b (right): Sagittal MPR image shows azygos continuation. The dilated azygos vein

(black arrow) drains instead of inferior vena cava (asterisk) ending at the top of the liver.

Ao; ascending aorta, LV; left ventricle

Figure 1a

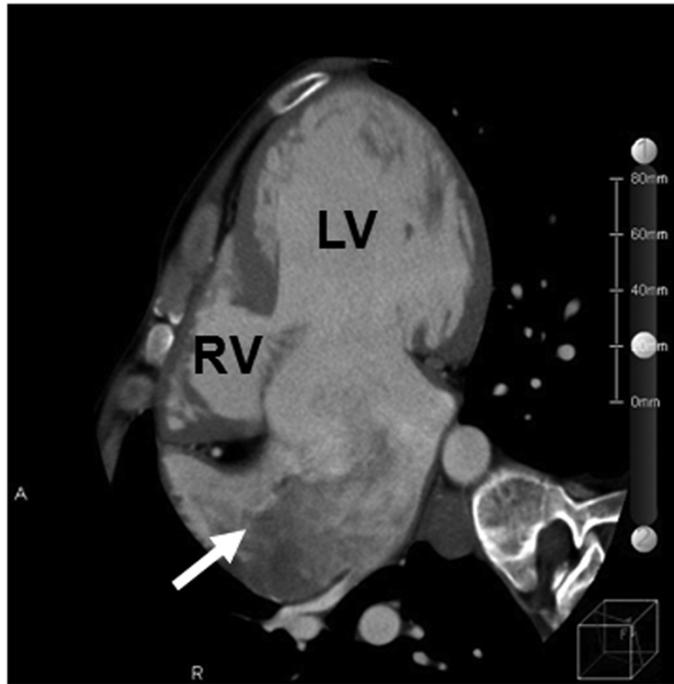


Figure 1b



Figure 2a

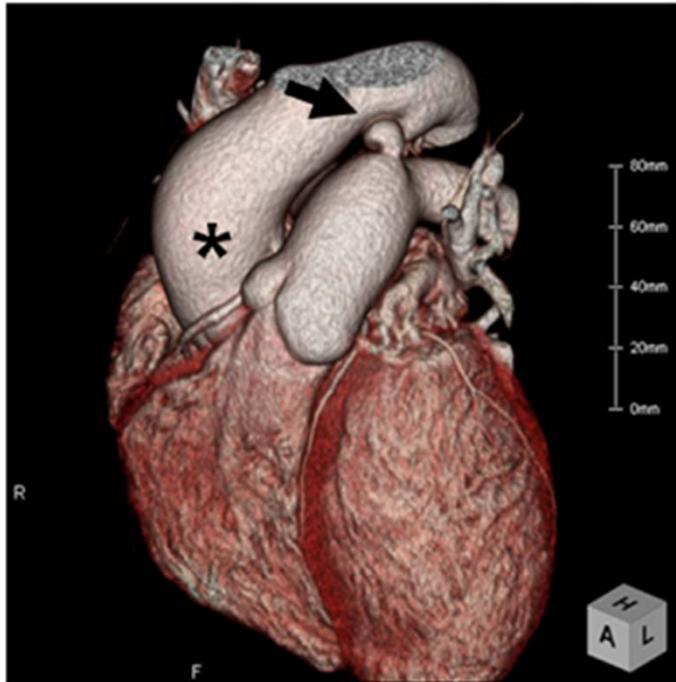


Figure 2b

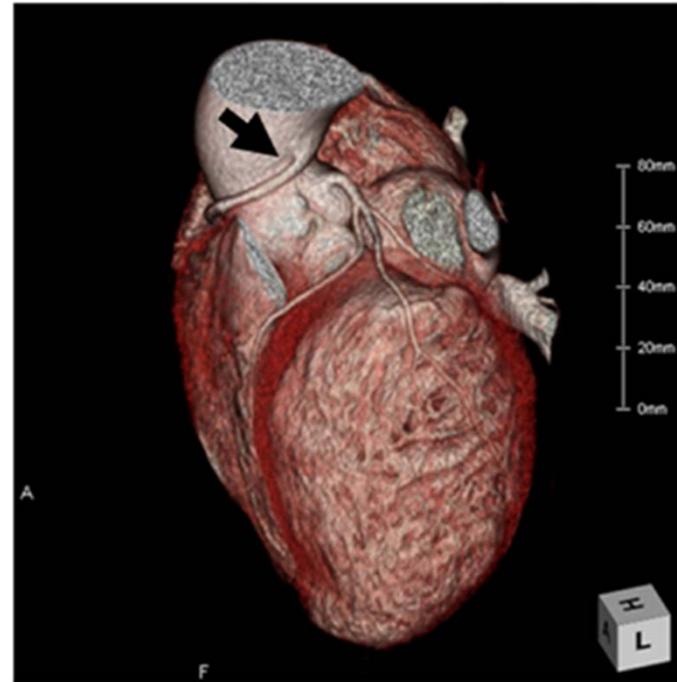


Figure 3a



Figure 3b

