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# Multidetector Computed Tomography Findings of Double-Outlet Right Ventricle Associated with Multiple Cardiac and Visceral Anomalies

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## ABSTRACT

A 25-year-old woman who had undergone the Blalock-Taussig shunt operation for double-outlet right ventricle (DORV) in her childhood was admitted to our hospital with mild cyanosis and dyspnea on exertion. To evaluate the precise complex anatomy of this abnormality, we carried out multidetector computed tomography (MDCT) angiography. MDCT clearly revealed both an occluded Blalock-Taussig shunt and a complex cardiac anatomy, including DORV, a doubly committed ventriculoseptal defect, pulmonary stenosis, persistent left superior vena cava, minor aortic arch anomalies, and total anomalous hepatic venous drainage. To our knowledge, our report is the first description of such a complex cardiac anatomy to be revealed with MDCT.

## INTRODUCTION

Double-outlet right ventricle (DORV) is a type of congenital cardiac anomaly in which both the aorta and the pulmonary artery originate from the right ventricle. Various anatomic subtypes exist, with a wide range of associated cardiac anomalies. Selection of the correct surgical technique is an important determinant of a patient's prognosis. Current imaging methods, including echocardiography, cardioangiography, and magnetic resonance imaging (MRI), have some drawbacks for delineating a complex cardiac anatomy. Multidetector computed tomography (MDCT) is a promising new technique for examination and follow-up of congenital heart disease. In this article, we described the MDCT findings of a case of DORV associated with multiple cardiac and visceral anomalies.

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## CASE REPORT

A 25-year-old woman who had undergone the Blalock-Taussig shunt operation in infancy was admitted to our hospital with mild cyanosis and dyspnea on exertion. A physical examination revealed cyanosis, clubbing, and grade 3/6 systolic murmur in the second left and right intercostal spaces. A chest radiograph showed mild cardiomegaly. An echocardiography evaluation revealed right ventricular hypertrophy, infundibulary pulmonary stenosis, and a muscular-type ventriculoseptal defect (VSD). A transthoracic echocardiographic examination demonstrated a hypoplastic left ventricle, a wide VSD, moderate pulmonary stenosis, and both great vessels (pulmonary artery and aorta) arising from the right ventricle.

To evaluate the precise complex anatomy of this abnormality, we performed MDCT angiography with a 64-slice multidetector row CT scanner (Brilliance 64 CT Scanner; Philips, Best, The Netherlands). We injected a 120-mL bolus of 60%-iodinated contrast material (iopamidol, Omnipaque 350 mg; GE Healthcare, Piscataway, NJ, USA) at a rate of 4 mL/s with a power injector. We used the bolus-tracking method with the following parameter values: 0.6 mm collimation, 12 mm per rotation table speed, and 0.5 mm increment, 120 kV, and 175 mA. All of the data were then transferred to a workstation (Extended Brilliance; Philips). Multiplanar reformatted views and volume-rendered and maximum-intensity projection images were obtained to analyze the complex anatomy. The MDCT examination confirmed that both great arteries originated from the morphologic right ventricle. There was a significant narrowing at the valvular level of the pulmonary artery, and the narrowing was mild in the infundibular area. The aorta was anterior to and to the right of the pulmonary artery (dextromalposition). The liver was at the left side of the abdomen, and the spleen and stomach were at the right side of abdomen owing to situs inversus (Figure 1). Two different VSDs were detected in multiplanar reformatted images: a doubly committed VSD related to both the aorta and the pulmonary artery, and a second noncommitted (membranous) VSD (Figure 2). The MDCT examination revealed both drainage of the hepatic veins directly into the left side of the left atrium and a persistent

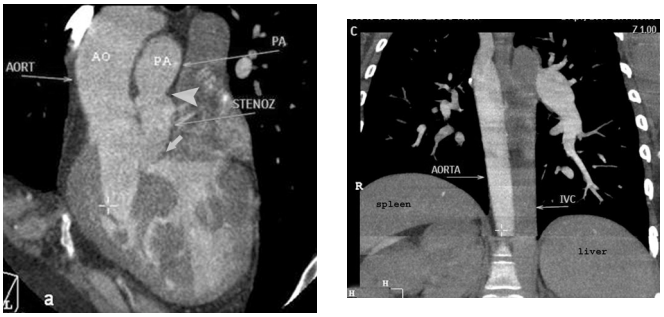


Figure 1. A, Oblique coronal multidetector computed tomography (MDCT) angiogram demonstrates both pulmonary artery (PA) and aortic root (AORT) arising from the right ventricle. There is narrowing at the valvular level of the PA (arrowhead), and narrowing is mild in the infundibular area (short thick arrow). STENOZ, stenosis. B, Coronal MDCT view showing the aorta anterior to and to the right of the PA (dextromalposition); the liver and the spleen are on the left and right sides of the abdomen, respectively, owing to situs inversus. IVC, inferior vena cava. C, A 3D volume-rendering MDCT image from anteroposterior view showing the aorta (star) anterior to and to the right of the pulmonary artery (arrow); both arise from the right ventricle. Arrowheads show the subclavian and left carotid arteries arising from arcus aorta.

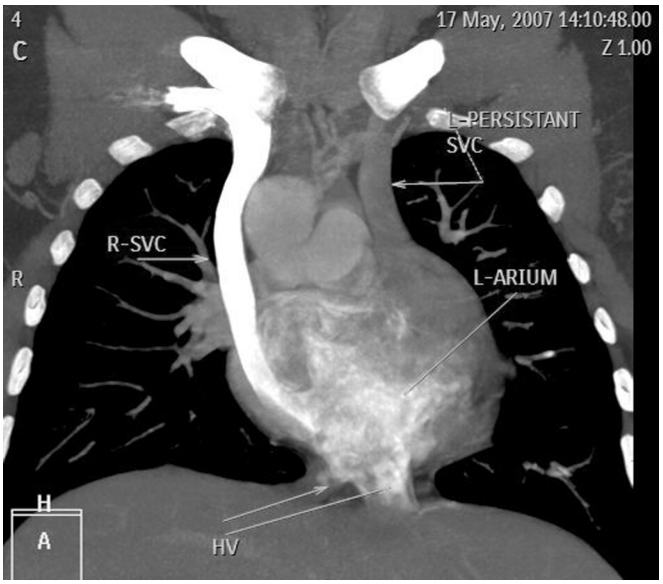


Figure 3. Multidetector computed tomography (MDCT) image demonstrating both of the hepatic veins (HV) draining directly into the left side of the left atrium and a persistent superior vena cava (SVC).

superior vena cava (Figure 3). Moreover, there were minor aortic arch anomalies (Figure 4). The Blalock-Taussig shunt was occluded (Figure 5). The coronary artery anatomy was normal (Figure 6).

The final MDCT diagnosis was an occluded Blalock-Taussig shunt, DORV, a doubly committed VSD, pulmonary stenosis, a persistent left superior vena cava, minor aortic arch anomalies, and a totally anomalous hepatic venous drainage.

## DISCUSSION

DORV requires at least 50% of each great vessel to arise from the morphologic right ventricle. A VSD is generally present in DORV, and its proximity to the arterial valves is the major determinant of the options for surgical treatment. VSDs in DORV have been classified into subaortic, subpulmonary, doubly committed, and noncommitted varieties according to the spatial relationship of the VSD to the arterial valves [Kirklin 1980]. The clinical signs and symptoms depend on both the associated VSD type and the additional cardiac

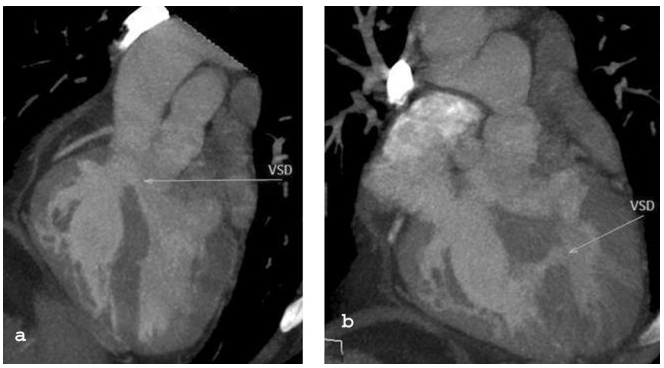


Figure 2. Oblique coronal multidetector computed tomography (MDCT) angiogram shows a doubly committed ventriculoseptal defect (VSD) (A) and a second, noncommitted membranous-type VSD (B).

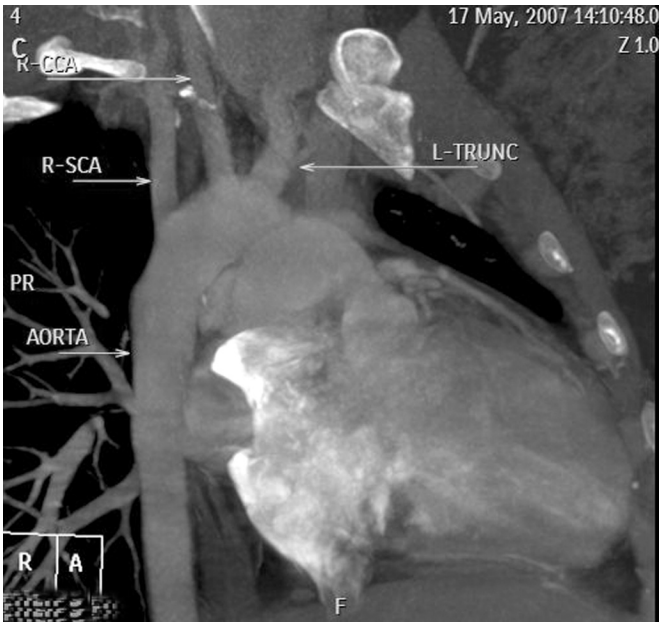


Figure 4. Oblique sagittal multidetector computed tomography (MDCT) angiogram showing right-sided aortic arch and its branch anomalies. R-SCA indicates right subclavian artery; R-CCA, right common carotid artery; L-Trunc, truncus brachiocephalicus.

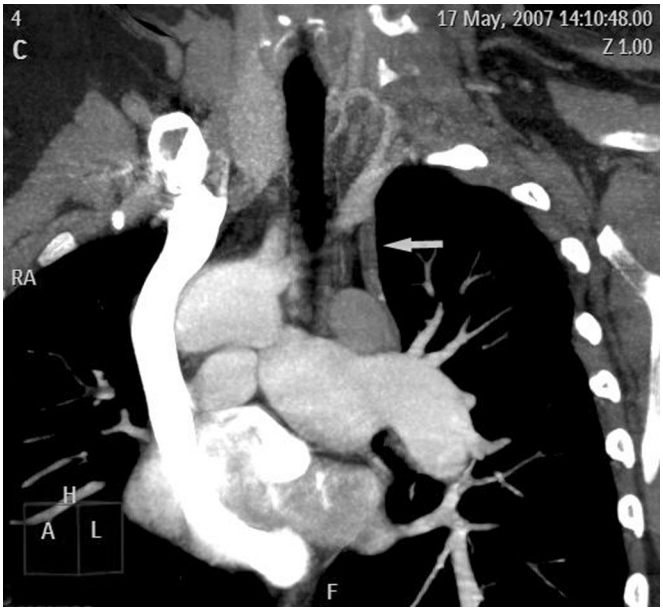


Figure 5. Oblique coronal multidetector computed tomography (MDCT) angiogram demonstrating the occluded Blalock-Taussig shunt (arrow).

anomalies. Various anatomic subtypes can be present. Many different types of either palliative or corrective surgical procedures have been developed [Kirklin 1980]. A careful description of the VSD and a determination of the relationship

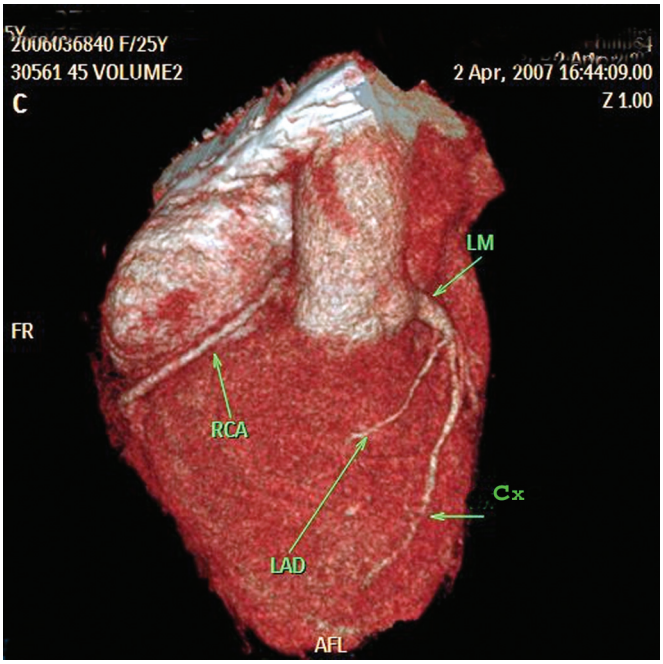


Figure 6. Volume-rendering image from the anterior view showing normal origins and courses of coronary arteries. RCA indicates right coronary artery; LM, left main coronary artery; LAD, left anterior descending artery; Cx, circumflex artery.

of the VSD with the semilunar valves, the relationship between the great arteries, and the situation of the right ventricular outflow tract are mandatory before surgical correction can be undertaken [Artrip 2006; Chen 2008].

Diagnosis of DORV can be made by conventional angiography, echocardiography, MDCT, and magnetic resonance angiography. Although echocardiography is considered the principal imaging technique, a comprehensive anatomic and functional analysis cannot be obtained with this method [Hartnell 1998; Kantarci 2007]. It is an operator-dependent method, and because of the limited sample size, it is not always reliable for identifying all features of VSD in DORV. Cinecardioangiography is an invasive technique and is not feasible for evaluating DORV and associated anomalies [Chen 2008]. MRI can depict the complex cardiac anatomy and associated lesions in almost all cases. It is an excellent adjunct for anatomic and functional evaluation, and the main advantages include a wide field of view and multiplanar capabilities. Nevertheless, longer scanning times (60–90 minutes) are needed for patients with complex congenital heart disease. The use of MRI is often limited in uncooperative patients [Didier 1986; Kantarci 2007]. MDCT has an expanding role in the comprehensive evaluation of congenital heart defects and postoperative follow-up. MDCT is not hampered by postoperative metal artifacts and is less time-consuming than MRI. The newer-generation MDCT scanners have better spatial resolution than MRI. A complex cardiac anatomy can be easily visualized with 3D/3D reconstructions [Ilica 2007].

MDCT can also be used to obtain functional data, including the ventricular ejection fraction, movement of the cardiac valves, and ventricular wall movement, and it is useful for morphologically evaluating rapidly moving intracardiac and paracardiac structures, including the coronary arteries [Pannu 2003; Ilica 2007; Kantarci 2007]. Chen et al [2008] reported the diagnostic accuracies of various methods for all VSD types in DORV (MDCT, 88%-100%; echocardiography, 71%-94%; cinecardioangiography, 60%-100%). MDCT allows a more complete evaluation of lung parenchyma, the mediastinum, and the upper abdomen. In our case, MDCT clearly revealed both an occluded Blalock-Taussig shunt and a complex cardiac anatomy, including DORV, VSDs, pulmonary stenosis, a persistent left superior vena cava, left atrial isomerism, minor aortic arch anomalies, and a totally anomalous hepatic venous drainage. One doubly committed VSD (related to both the aorta and the pulmonary artery) and a second, noncommitted (membranous) VSD were accurately demonstrated with multiplanar CT images.

MDCT is able to recognize the type of congenital variation and the configuration of additional malformations to facilitate accurate diagnosis and sufficient treatment. A more accurate presurgical diagnosis with the use of MDCT may improve surgical prognosis and decrease mortality rates. Moreover, MDCT can reveal many other associated cardiac and visceral anomalies. In conclusion, MDCT is a candidate method for "one-stop-shop" evaluation of congenital heart disease in the near future.

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