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CONGENITALLY ABSENT RIGHT CORONARY ARTERY IN ADULT

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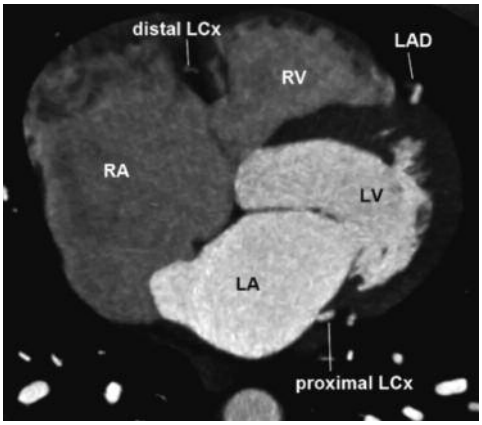
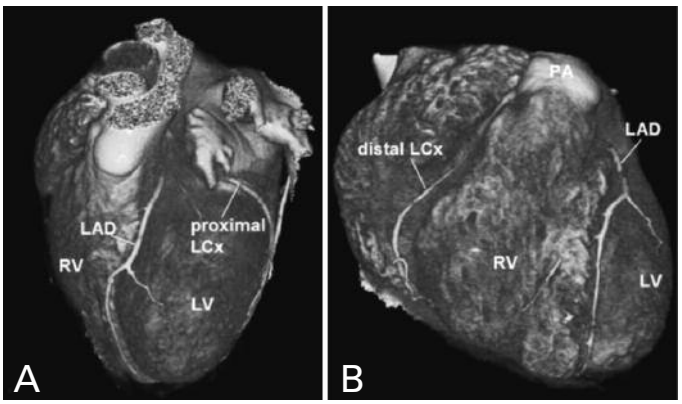
Key-word: Coronary arteries, abnormalities

Background: A 44-year-old male patient with atrial fibrillation and cardiac insufficiency (NYHA IV) presented for evaluation for heart transplantation. He was referred for cardiac CT for assessment of potential restrictive cardiomyopathy and evaluation of his right ventricle. Cardiac CT was performed utilizing a dual-source CT scanner. ECG-adapted tube current was used to reduce radiation exposure. No pericardial calcifications were noted. The right atrium and the inferior vena cava were dilated with 82 x 58 mm and 47 mm diameter, respectively. There was no evidence of coronary artery disease.





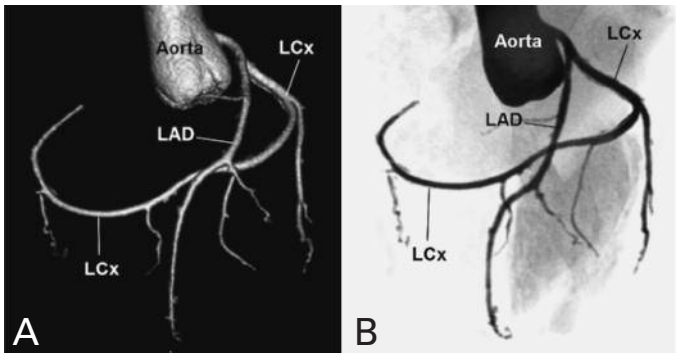


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Fig.	2A	2B
	3A	3B

Work-up

Transversal thin maximum intensity projection (thin MIP) of contrast-enhanced coronary CT (Fig. 1) demonstrates normal caliber of the left anterior descending artery and proximal left circumflex artery, and small caliber of the distal LCx in the right atrioventricular grove. The right coronary artery is missing.

3D volume rendered technique (VRT) of contrast-enhanced cardiac CT (Fig. 2) shows normal course of the LAD. The LCx is elongated and bends around the heart into the right atrioventricular grove, a vascular territory that normally is supplied by the right coronary artery. Please note the regular diameter of the proximal LCx as compared to the small lumen of the distal LCx within the right atrioventricular grove.

VRT (Fig. 3A) and inverted angio view of the same 3D data set (Fig. 3B) display the coronary artery tree. The left circumflex artery is elongated and reaches into the territory of the absent right coronary artery. No coronary artery stenosis is present. (LAD = left anterior descending artery, LCx = left circumflex artery, RV = right ventricle, LV = left ventricle, RA = right atrium, LA = left atrium).

Radiological diagnosis

The CT-findings reveal an explicitly rare *coronary artery anomaly* consisting of entirely absent right coronary artery. Instead, the left circumflex artery (LCx) is elongated and has a course around the apex of the heart into the right atrioventricular grove.

The lumen of the LCx and its marginal branches is patent. The left main and the left anterior descending artery (LAD) with its diagonal branches are normal.

Discussion

Coronary artery anomalies are a relatively rare finding. However, with increasing use of non invasive imaging modalities such as MRI and CT depiction of coronary anomalies becomes more frequent and the incidence is reported to be as high as 5.6%. An ectopic RCA, arising from right sinus or the left sinus are the most common coronary anomalies with an incidence of 1.13% and 0.92%, respectively. In a large study of 1950 angiograms an absent left main artery and left circumflex artery arising from the right sinus were the next most common coronary anomalies with an incidence of 0.67% each.

Absence of the right coronary artery is an explicitly rare finding. In a large analysis of 58 023 coronary angiographies only two cases (0.0034%) of absent RCA were detected. However, its true incidence in the general population is unknown.

The clinical presentation of coronary artery anomalies may vary from entirely asymptomatic patients to exertional angina, myocardial ischemia or even sudden cardiac death. Coronary artery anomalies with an interarterial course, i.e. between the pulmonary artery and the ascending aorta, are thought to be most hemodynamically significant.

Conventional coronary angiography may display the presence of a coronary anomaly. However, for defining the ostial origin and exact course of anomalous coronary arteries multidetector ECG-gated CTA has been shown to be superior to angiography. Coronary MR-angiography (MRA) may also be used to noninvasively identify coronary artery anomalies, without the use of ionizing radiation or contrast agent. Both CTA and MRA are noninvasive diagnostic modalities that eliminate the risks inherent in invasive angiography while covering an entire volume of interest in high spatial and temporal resolution.

This report illuminates the benefit of noninvasive coronary CTA for assessment of coronary anomalies in a rare case of an absent right coronary artery with an elongated left circumflex artery. Coronary artery and pericardial disease could be excluded and the exact course of the anomalous left circumflex could readily be revealed.

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