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Congenital Hypoplastic Left Coronary Cusp

A 54-year-old male presented to our emergency department with chest tightness and chest pain for 5 h. Physical examination was unremarkable. Laboratory tests showed elevated cardiac troponin levels with hs-cTnI 4.238 ng/L (normal value <0.0198 ng/L). Echocardiography showed ascending aortic dilatation, severe aortic valve insufficiency, and left ventricular wall thickening. Coronary computed tomographic angiography (CCTA) showed a hypoplastic left coronary cusp separated by membranous structure, and agric valve leaflet thickness with calcification (Figures 1A and B); the left anterior descending and the left circumflex coronary artery were hypoplastic; left sinus of Valsalva and the left coronary artery (LCA) were perfused by multiple collaterals from the dominant right coronary artery (RCA) (Figure 1C). Selective RCA angiography confirmed normal RCA with a well-developed collateral flow to the LCA; but the ostium of the LCA was patent, and the left sinus of Valsalva was filled with retrograde LCA blood flow (Figure 1D, Video 1). He was recommended for surgical aortic valve replacement and excision of the rudimentary cusp, but the patient refused and was discharged.

C D LCX LCX LAD

Figure 1. (A and B) Multiplanar reconstructions computed tomography angiography image shows a pouch-like filling in the small left coronary aortic sinus with the obstruction of the left main coronary ostium. (C) The coronary tree image shows that the LAD and the LCX were hypoplastic; left sinus of Valsalva and the left coronary artery were perfused by multiple collaterals from the dominant (RCA). (D) Selective RCA angiography confirmed normal RCA with a well-developed collateral flow to the LCA and the left sinus of Valsalva was filled with retrograde LCA blood flow. LAD, left anterior descending; LCA, left coronary artery; LCX, left circumflex; RCA, right coronary artery.



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E-PAGE ORIGINAL IMAGE





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Congenital hypoplastic left coronary cusp separated by a membrane-like structure is an extremely rare but noteworthy anomaly. This leads to a progressive obstruction of blood flow to the LCA, causing recurrent acute myocardial infarction and sudden death. The typical characteristic of coronary angiography and CCTA is a pouch-like filling in the small left sinus of Valsalva, which is an occlusion of the coronary ostium by a rudimentary aortic cusp. Patients may experience symptoms of myocardial ischemia, and surgical repair of the coronary ostium is often the only way to treat this anomaly.

Informed Consent: The informed consent was obtained from the patient for this study.

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Video 1: Transcatheter coronary angiography shows a normal RCA with a well-developed collateral flow to the LCA, and the left sinus of Valsalva is pouch-like, filling with retrograde LCA blood flow. LAD, left anterior descending; RCA, right coronary artery; LCX, left circumflex.

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