

## Percutaneous treatment of huge congenital coronary-cameral fistula

### Dev koroner-odacık fistülünün perkütan tedavisi

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**Summary**– An 11-year-old girl was admitted to our hospital with a history of fatigue, effort dyspnea, and chest pain. On transthoracic echocardiography, the parasternal short-axis and apical four-chamber color Doppler flow imaging showed a structure matching the level of the coronary sinus and pouring into the right ventricle. On multi-slice computed tomography, the circumflex artery became immediately narrowed just before draining in to the base of the right ventricle, close to the septal leaflet of the tricuspid leaflet, and the Qp/Qs was 1.6. Congenital coronary-cameral fistula is an infrequent condition that establishes a direct link between coronary arteries and cardiac chambers. Although coronary artery fistulae are commonly asymptomatic, they may cause severe symptoms depending on the severity of the shunt, and may be treated medically, surgically or by transcatheter closure. In this case report, we present a patient with a huge circumflex artery-to-right ventricle fistula treated successfully with a duct occluder device.

Coronary-cameral fistula (CCF) is an abnormal communication arising from one or more coronary arteries and draining into one or more heart chambers. The vast majority of cameral fistulae communicate with the right-sided chambers of the heart, and in the remainder of cases will drain to the left side of the heart or to both sides.<sup>[1]</sup> Fistulae usually arise predominantly from the right coronary artery (RCA); however, in a small proportion of cases, communications may arise from both the right and left coronary tree.<sup>[2]</sup> Two cases have been reported in

**Özet**– On bir yaşındaki kız çocuğu halsizlik, efor dispnesi ve göğüs ağrısı şikayetleri ile hastanemize başvurdu. Transtorasik ekokardiyografide, parasternal kısa aks ve apikal dört boşluk renkli Doppler akım incelemelerinde koroner sinüs seviyesinde sağ ventriküle dökülen bir yapı izlendi. Çok kesitli bilgisayarlı tomografide bu yapının sirkumfleks arter olduğu ve triküspit septal kapakçığı komşuluğunda sağ ventriküle döküldüğü görüldü. Hesaplanan Qp/Qs değeri 1.6 idi. Koroner arterlerle kalp odacıkları arasında bağlantı olan doğumsal koroner-odacık fistülleri oldukça nadir durumdur. Koroner arter fistülü genellikle semptomsuz olmasına rağmen şantın ciddiyetine bağlı olarak ciddi semptomlara da neden olabilirler ve tıbbi, cerrahi ya da transkateter yolla tedavi edilebilirler. Bu olgu bildirisinde, sirkumfleks arterden sağ ventriküle doğru olan fistülün dukt kapama cihazı ile başarılı bir şekilde tedavi edildiği hasta sunuldu.

which the RCA was connected to the right ventricle (RV), and both underwent successful percutaneous closure.<sup>[3,4]</sup>

In this report, we present a patient with a huge circumflex artery-to-RV fistula treated successfully with a duct occluder device.

#### Abbreviations:

CCF	Coronary-cameral fistula
Cx	Circumflex
LCx	Left circumflex
LMCA	Left main coronary artery
MSCT	Multi-slice computed tomography
PDA	Patent ductus arteriosus
RCA	Right coronary arteries
RV	Right ventricle

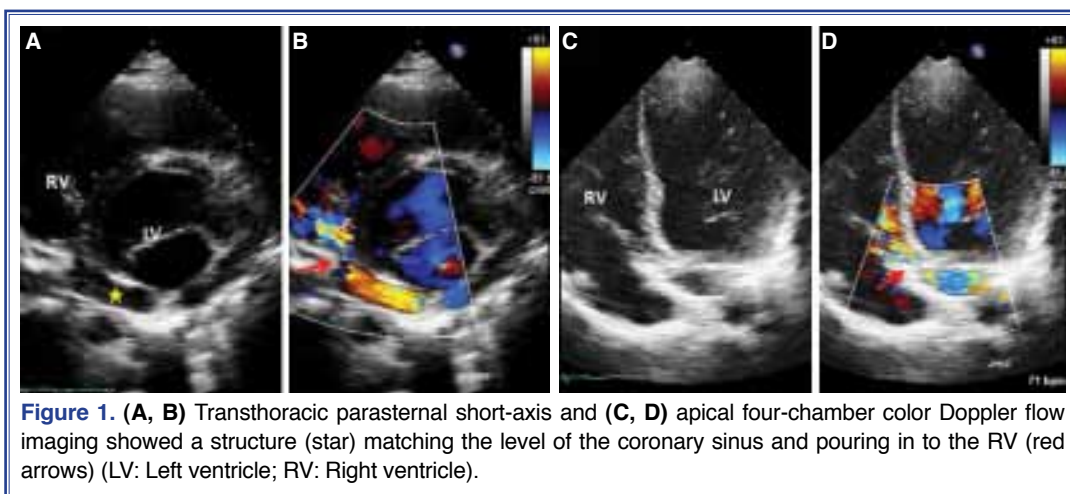
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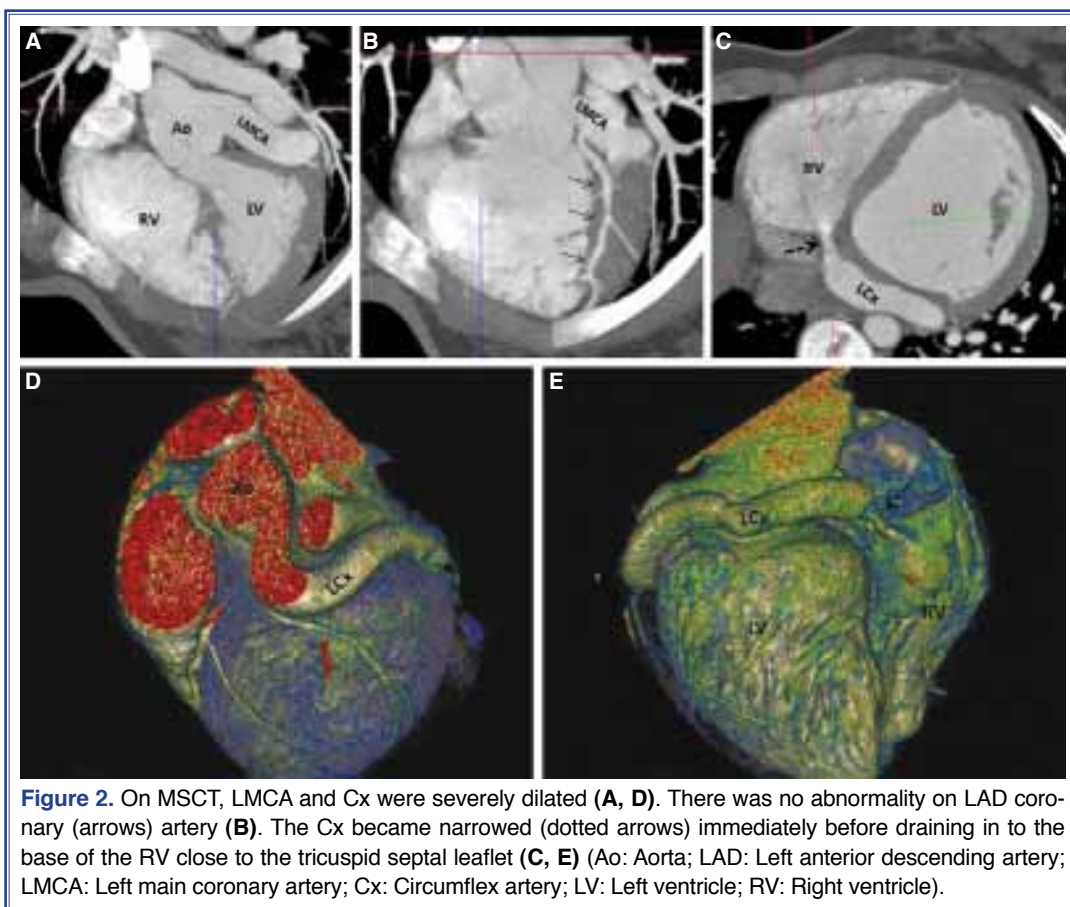




### CASE REPORT

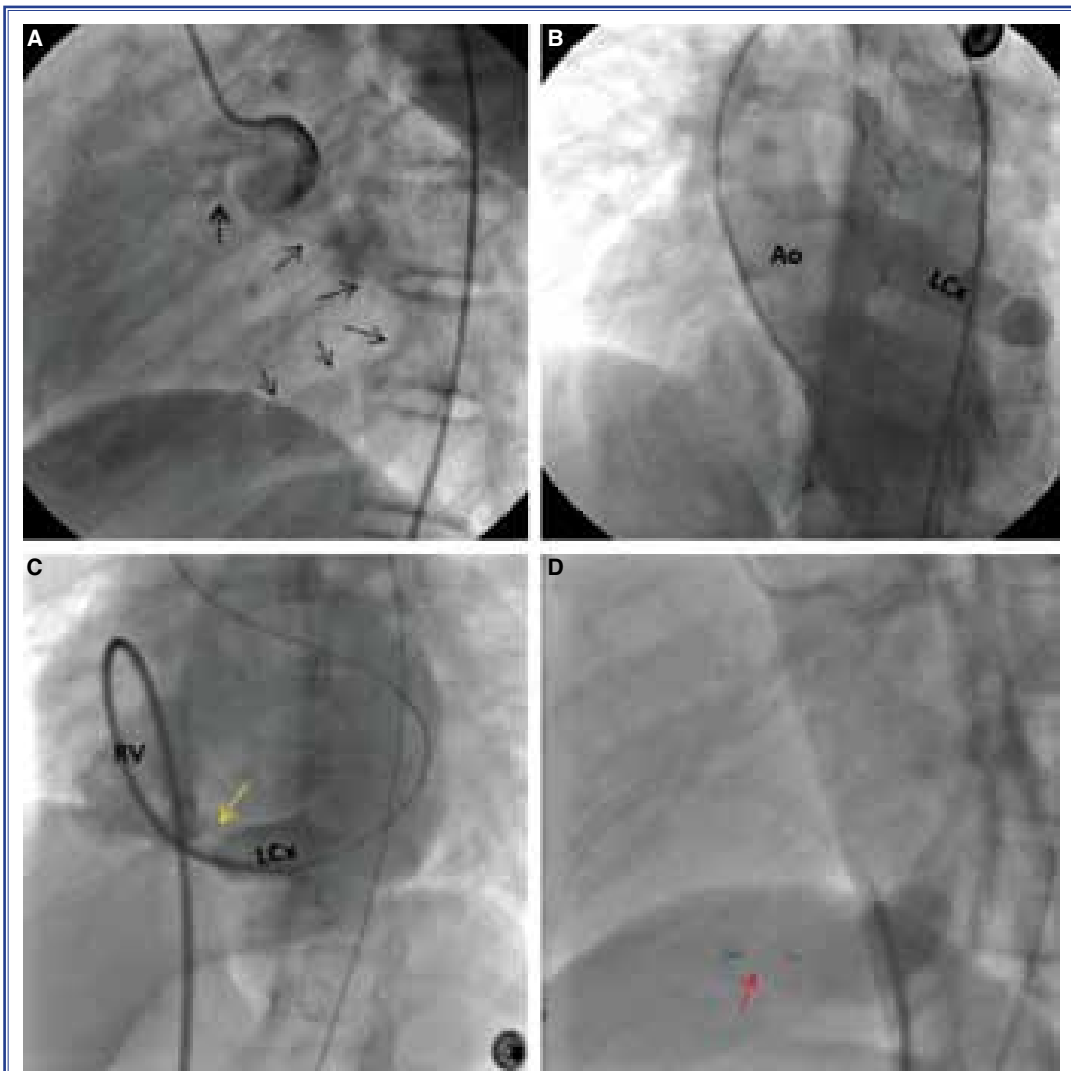
An 11-year-old girl was admitted to our hospital with a two-year history of fatigue, effort dyspnea, and chest pain. On the physical examination, there was a continuous grade 2-3 murmur best heard at the

lower left parasternal area. The ECG showed normal sinus rhythm. On transthoracic echocardiography, the RV was dilated and pulmonary artery pressure was elevated. In addition, the parasternal short-axis and apical four-chamber color Doppler flow imaging showed a structure matching the level of the

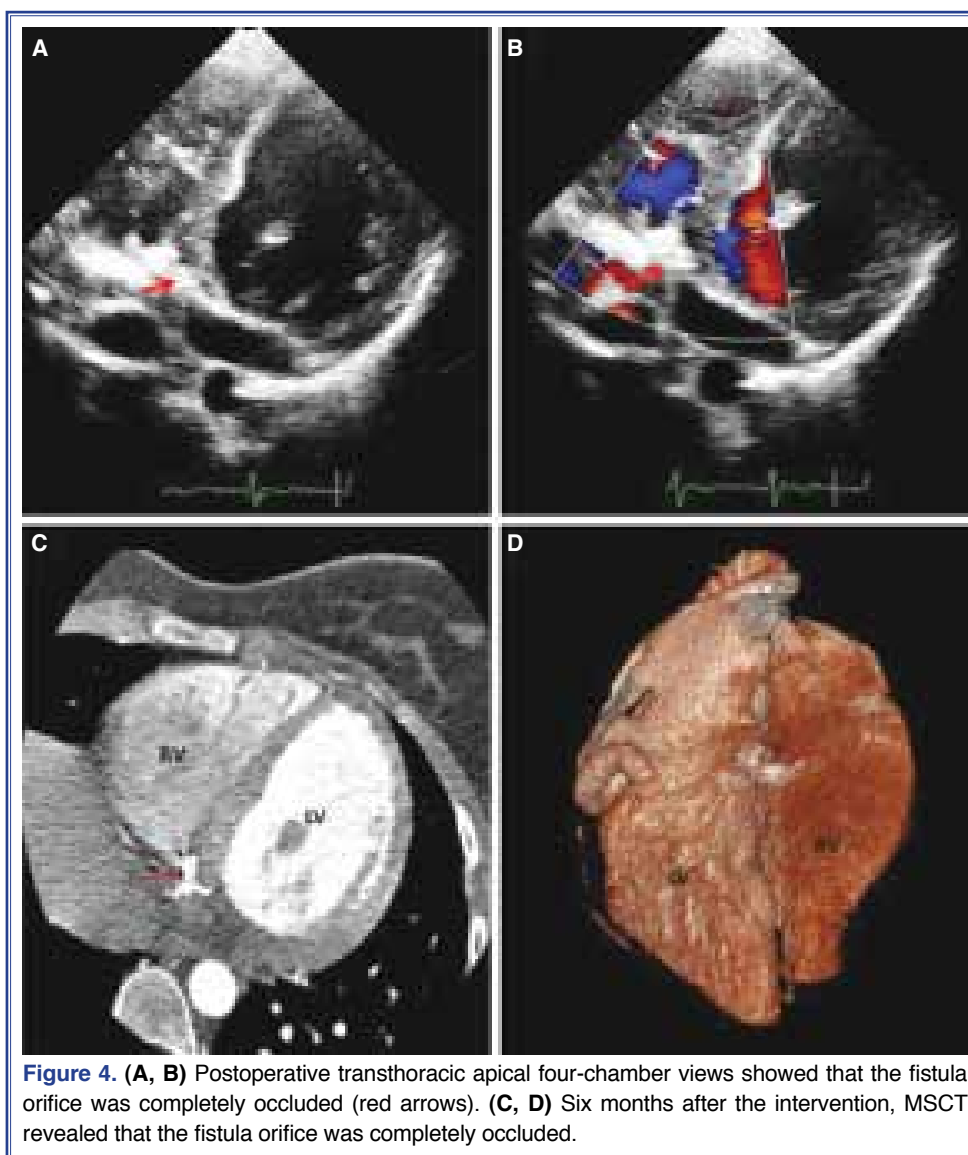


coronary sinus and pouring in to the RV (Figure 1 and Video 1\*). On multi-slice computed tomography (MSCT), the left main (LMCA) and circumflex (Cx) arteries were severely dilated. It clearly seemed that the structure seen on echocardiography was a severely dilated Cx. There was no anomaly on the left anterior descending artery (LAD) and RCA (Figure 2). The Cx became narrowed immediately before draining in to the base of the RV close to the septal leaflet of the tricuspid leaflet. The narrow segment of

the fistula was 2.5 cm in length, and its diameter was 0.61 cm at the proximal end and 0.71 cm at the fistula orifice. After consultation with the family about surgical or percutaneous intervention options, coronary intervention was decided. On cardiac catheterization, systolic/diastolic/mean pulmonary arterial pressures were 42/18/30 mmHg, respectively, and Qp/Qs was 1.6. Aortic root angiography demonstrated the huge aneurysmal fistula beginning as LMCA, continuing as left Cx and draining into the RV. Contour of



**Figure 3.** (A) Left coronary angiography and (B) left ventriculography demonstrated the huge aneurysmal fistula beginning as LMCA, continuing as Cx and draining into the RV. (C, D) After a 9F destination sheath was introduced through the femoral vein and advanced to the narrowest segment of the fistula (yellow arrow), a PDA occluder device (red arrow) was released into the fistula. Final left coronary angiography showed that there was no contrast medium flowing into the RV through the fistula orifice (Arrows and dotted arrow show Cx and LAD arteries, respectively) (Ao: Aorta; RV: Right ventricle; LAD: Left anterior descending artery; LCx: Left circumflex artery).



**Figure 4.** (A, B) Postoperative transthoracic apical four-chamber views showed that the fistula orifice was completely occluded (red arrows). (C, D) Six months after the intervention, MSCT revealed that the fistula orifice was completely occluded.

the fistula was smooth, and no major distal branches near the termination were seen. Contrast agent poured into the RV from the LCx, and the fistula orifice was about 0.9 cm. On coronary sinus angiography, there was no relationship between the fistula and coronary sinus. Vascular access was attained via the right femoral artery and left femoral vein using 6F and 8F angiography catheters, respectively. A 0.028 hydrophilic guide wire was advanced through the Judkins left #5 coronary arteriography catheter along the fistula until reaching the main pulmonary artery; the wire was snared by a gooseneck catheter and an arteriovenous loop was created (Figure 3 and Video 2\*). Then, the hydrophilic guide wire was re-

moved and a 0.35 stiff guide wire was placed through a multipurpose 6F catheter. After a 9F destination sheath was introduced through the femoral vein and advanced to the narrowest segment of the CCF, a patent ductus arteriosus (PDA) occluder device (8 x 10 mm) was released into the fistula. After delivery of the occluder, the angiography showed that there was no contrast medium flowing into the RV through the fistula orifice. The patient was discharged on clopidogrel and warfarin therapy. Six months after the intervention, echocardiography and MSCT revealed that the fistula orifice was completely occluded and there was no leak in to the RV (Figure 4). Based on MSCT findings, it was decided to withdraw clopido-

grel, but continue warfarin therapy. The patient remains on warfarin therapy.

## DISCUSSION

Coronary-cameral fistula (CCF) is an abnormal communication arising from one or more coronary arteries and draining into one or more heart chambers. The vast majority of cameral fistulae communicate with the right-sided chambers of the heart, and in the remainder of cases, will drain to the left side of the heart or to both sides.<sup>[1]</sup> Fistulae usually arise predominantly from RCA; however, in a small proportion of cases, communications may arise from both the right and left coronary tree.<sup>[2]</sup> Two cases were reported with RCA connecting to the RV, which were successfully closed percutaneously.<sup>[3,4]</sup> Clinical presentations will generally depend on the hemodynamic significance of the anomaly. Although coronary artery fistulae are most commonly asymptomatic and found incidentally, they may cause symptoms of angina pectoris. Further, CCF may cause myocardial infarction, congestive heart failure, arrhythmias, aneurysmal formation, and rupture of affected vessels.<sup>[5-8]</sup> The best way to manage CCF is uncertain largely due to the rarity of this pathology. Patients in whom fistulae with large shunts exist may benefit from closure of the shunt, and it is probably best to be done as early as possible. Management of a CCF depends on the experience of each medical center; valid options include primary surgical treatment or transcatheter closure. Prior to the availability of occlusive devices, surgery was the only choice. While the reported surgical morbidity and mortality and incidence of recurrence are low, such procedures still require a median sternotomy and usually cardiopulmonary bypass.<sup>[9]</sup> Catheter closure techniques have been performed to treat coronary fistulae with several devices, including detachable balloons, stainless steel coils, controlled-release coils, controlled-release PDA coils, and Amplatzer PDA plug.<sup>[10-13]</sup>

The operator must be aware of embolization while performing transcatheter closure, especially in fistulae arising from the LMCA. The complication of coil occlusion is rare but may include coronary artery occlusion due to thrombosis. There are cases reported in the literature in which low-dose aspirin and warfarin addition for prophylaxis of thrombosis were used if the coronary dilatation was severe (>10 mm),

particularly when coronary flow is sluggish.<sup>[14]</sup> However, there is a little information available regarding the risk of coronary thrombosis in this group, and the benefits are controversial.

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**\*Supplementary video files associated with this article can be found in the online version of the journal.**

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**Key words:** Cardiac catheterization; child; coronary angiography; coronary vessel anomalies; echocardiography, Doppler, color; percutaneous coronary intervention; vascular fistula.

**Anahtar sözcükler:** Kalp kateterizasyonu; çocuk; koroner anjiyografi; koroner damar anomalisi; ekokardiyografi, Doppler, renkli; perkütan koroner girişim; vasküler fistül.