

A rare coronary anomaly with masked diagnosis: Anomalous left circumflex artery from right pulmonary artery

Tanı konulamamış nadir bir koroner anomali: Sağ pulmoner arterden çıkan sirkumfleks arter

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Summary– Anomalous origin of the circumflex coronary artery from the pulmonary artery is a rare congenital coronary anomaly. While it generally follows an asymptomatic course, if undiagnosed it may lead to severe clinical outcomes, including sudden death. The condition can be masked by associated defects, so when it is clinically suspected, diagnosis must be confirmed by conventional and/or magnetic resonance angiography, even if echocardiography clearly shows coronary roots. This report describes a patient who underwent neonatal surgery for aortic coarctation and was diagnosed with coronary artery anomaly at 15 months old.

Özet– Sirkumfleks koroner arterin pulmoner arterden köken alması nadir görülen bir doğumsal koroner arter çıkış anomalisidir. Genellikle semptomsuz seyretmekle birlikte tanı konulamaması durumunda nadiren ciddi klinik bulgulara ve hatta ani ölüme yol açabilir. Tanı koydurucu bulgular eşlik eden lezyonlar nedeni ile maskelenebileceğinden ekokardiyografik değerlendirmede koroner kökler net izlense de klinik şüphe varlığında anjiyografi ve/veya manyetik rezonans anjiyografi ile tanı kesinleştirilmelidir. Bu yazıda, yenidoğan döneminde aort koarktasyonu nedeni ile ameliyat edilen ve takiplerinde 15 aylık iken koroner arter çıkış anomalisi tanısı konulan olguyu sunuyoruz.

Coronary artery origin anomalies are rare, occurring as an isolated anomaly in 0.3–0.9% of the population, and accompanied by other congenital cardiovascular defects in 3–36%.^[1] Of these anomalies, anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is one of the most serious, but also one of the most well-researched. It can have a loud clinical presentation and lead to dilated cardiomyopathy and congestive heart failure in the early infant period.^[2] Anomalous right coronary

Abbreviations:

ACCAPA	Anomalous circumflex coronary artery from pulmonary artery
ALCAPA	Anomalous left coronary artery from pulmonary artery
ARCAPA	Anomalous right coronary artery from pulmonary artery
LAD	Left anterior descending
LCA	Left coronary artery
LCX	Left circumflex artery
PDA	Patent ductus arteriosus
RCA	Right coronary artery
RPA	Right pulmonary artery
VSDs	Ventricular septal defects

or circumflex coronary arteries from pulmonary artery (ARCAPA and ACCAPA respectively) are very rare in comparison. They often occur together with other congenital cardiac pathologies and are either asymptomatic or have a milder clinical presentation than ALCAPA.^[3]

This report describes a patient who underwent neonatal surgery for aortic coarctation.

CASE REPORT

A 15-month-old male patient with discrete aortic coarctation, a patent ductus arteriosus (PDA), mild mitral stenosis (peak 8 mmHg, mean 4 mmHg gradient), multiple small muscular ventricular septal defects (VSDs), left ventricular hypertrophy and pulmonary hypertension underwent surgery in the neonatal period and showed no clinically significant signs in post-

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operative follow-up. Coronary artery anomaly was not suspected in the pre-operative period. However, post-operative echocardiographic examination in the first year of life demonstrated a dilated left coronary artery despite the absence of systolic dysfunction, mitral valve regurgitation or a dilated left ventricle. Color flow imaging also showed multiple small muscular VSDs in the ventricular septum or the candle flame appearance, which could be interpreted as coronary collaterals, as well as diastolic flow in the right pulmonary artery (RPA) (Video 1, 2*). As these findings pointed to a possible coronary fistula or ACCAPA, angiography was planned. In addition to the echocardiographic findings listed above, the mild mitral valve stenosis detected in the neonatal period still persisted.

Pre-operative and post-operative ECG findings were not suggestive of ischemia. Angiography showed pulmonary artery pressure of 30/15 (19) mmHg, and ascending aorta pressure of 94/51 (64) mmHg. There was no residual gradient between the ascending and the descending aorta. Left ventricle injection showed no VSD. Selective injection into the right and left coronary artery (RCA, LCA) demonstrated origin of the RCA and the left anterior descending artery (LAD) from the aortic root. It was also observed that the left circumflex artery (LCx) did not arise from the

left coronary system, but was retrogradely filled by multiple collaterals from the RCA and LCA during the late phase. Diastolic flow was also observed in the RPA (Figure 1, Video 3 and 4*). It was decided to re-implant the circumflex artery to the aortic root and close the pulmonary artery opening with a pericardial autograft.

DISCUSSION

While asymptomatic in some cases, coronary artery anomalies may lead to myocardial infarction, arrhythmia, ventricular aneurysm, mitral regurgitation, cardiomyopathy and sudden death.^[4] ACCAPA is a very rare coronary anomaly and is often accompanied by other pathologies,^[5] the most common of which are aortic coarctation, PDA, Tetralogy of Fallot, aortopulmonary window, truncus arteriosus, VSD and pulmonary valve stenosis.^[6] Our patient had aortic coarctation, PDA and mild mitral valve stenosis.

Judging from the limited number of ACCAPA cases reported in the literature, the clinical presentation is highly variable.^[3] Some cases are symptomatic in the early infant period, while some are asymptomatic until adulthood.^[3,7] The clinical presentation likely depends on how developed the collateral vessels are.^[8] Our patient was asymptomatic with no coronary findings, and the anomaly was first suspected during echocardiographic examination.

While typical ECG findings in symptomatic patients are well-documented, they may be unremarkable in asymptomatic patients.^[9] In our patient, no complaints or symptoms were observed throughout the post-operative follow-up period, and ECG was unremarkable except for incomplete right bundle branch block.

Echocardiography may be insufficient for diagnosis of ACCAPA, which can be clinically masked by concomitant pathologies.^[5] Although in most reported cases echocardiography rarely shows clear coronary anatomy findings, symptomatic patients demonstrate disrupted contractility and low ejection fraction.^[2] In some reports, echocardiographic examination before diagnosis clearly showed RCA and LCA origins and dilated coronary vessels. This has been attributed to the compensatory mechanism of coronary steal syndrome caused by collateral vessels.^[5] In patients with a more severe clinical presentation and congestive heart failure, echocardiographic findings are left

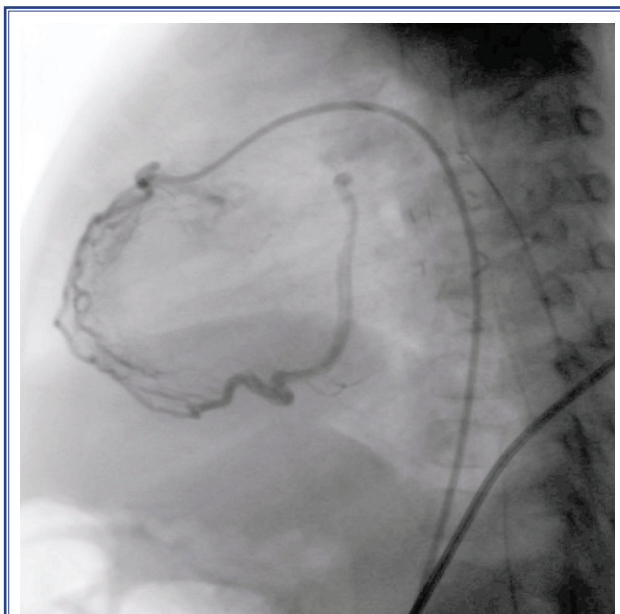


Figure 1. Coronary angiography shows the retrograde filling of an anomalous left circumflex artery through collateral vessels provided by the left anterior descending and the right coronary artery on angiography.

ventricular dilation and major disruption of cardiac function.^[10] Similarly, our patient had normal RCA and LCA origins in the pre- and post-operative period. Despite normal appearance of LCA origin, if this artery is dilated and coronary bifurcation cannot be visualized for a long segment in addition to other findings, LCx or LAD origin anomaly must be considered. Due to the concomitant pathologies such as aortic coarctation, PDA, mild mitral valve stenosis, left ventricular hypertrophy and pulmonary hypertension, we did not at first see the mild coronary dilation and coronary flow into pulmonary artery. That is, the flow from the LCx into the RPA was not observed early on. In follow-up, coronary vessels were comparatively dilated, and the appearance of multiple muscular VSDs in the interventricular septum—interpreted to exist as the result of collaterals—and diastolic flow in the RPA pointed to a possible congenital coronary anomaly and angiography was performed to confirm the diagnosis.

Angiography is the gold standard in diagnosis of ACCAPA. According to the literature, magnetic resonance and conventional angiography are equally effective and the latter is not obligatory in every case.^[11] Conventional angiography with aortic root and selective coronary injections allows for clear visualization of the coronary anatomy and the collateral vessels. In our case, angiography confirmed the presence of collaterals arising from the RCA and LAD that retrogradely filled the LCx and pulmonary artery. Contrast injection into the left ventricle showed no VSD. While in most patients ACCAPA arises from the RPA, there have also been cases with origin from the LPA and main pulmonary artery.^[2] Angiographic examination in our patient showed that the LCx arose from the RPA.

ACCAPA requires surgical treatment. While ligation and bypass grafting are recommended in adults,^[12] re-implantation yields substantially better results in infants and children.^[10] We re-implanted the circumflex artery to the aortic root and closed the pulmonary artery opening with a pericardial autograft.

ACCAPA is a rare congenital cardiac anomaly which, if undiagnosed, that can lead to severe clinical findings and sudden death. As the condition can be masked by its associated defects, in a case of clinical suspicion, and even if echocardiography clearly shows coronary roots, diagnosis must be confirmed by conventional and/or magnetic resonance angiography.

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

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