Evaluation of patients with coronary artery anomalies

Koroner arter anomali olan hastaların değerlendirilmesi

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ABSTRACT

Background: This study aims to evaluate patients with coronary artery fistulas, anomalous origin of left coronary from pulmonary artery, and anomalous origin of right coronary artery from pulmonary artery.

Methods: Fourteen patients who were referred to pediatric cardiology clinic with a coronary artery fistula, anomalous origin of left coronary from pulmonary artery, and anomalous origin of right coronary artery from pulmonary artery between January 2009 and March 2015 were retrospectively analyzed. Ten patients with a coronary artery fistula were grouped as group 1, and four patients with anomalous origin of left coronary from pulmonary artery and anomalous origin of right coronary artery from pulmonary artery as group 2. Most of the patients were evaluated for chest pain with murmur on physical examination. Three patients underwent transcatheter and one patient surgical procedure. One patient indicated for surgery did not accept treatment and was followed in the outpatient setting with the other five patients who did not need any intervention. Patients in group 2 underwent surgery.

Results: The patients who underwent treatment were discharged from hospital without any problem. Acetylsalicylic acid was given to the patients in group 1 following the intervention. One patient from hospital without any problem. Acetylsalicylic acid was given also warfarin for persistent dilatation of the proximal right coronary artery. Group 2 patients had improved clinical findings during follow-up.

Conclusion: Coronary artery fistulas, anomalous origin of left coronary from pulmonary artery, and anomalous origin of right coronary artery from pulmonary artery can be easily overlooked during echocardiographic evaluation. Therefore, further imaging modalities may be needed. Transcatheter approach is a safe alternative to surgical therapy in the treatment of fistulas. Arterial reimplantation technique is successful in anomalous origin of left coronary artery from pulmonary artery and anomalous origin of right coronary artery from pulmonary artery surgery. Echocardiography has the main role in identifying these rare, but critical conditions.

Keywords: Anomalous origin of coronary artery; coronary artery fistula; echocardiography.

ÖZ

Amaç: Bu çalışmada koroner arter fistülleri, pulmoner arterden sol koroner arterin anormal çıkışı ve pulmoner arterden sağ koroner arterin anormal çıkışı olan hastalar değerlendirildi.


Anahtar sözcükler: Koroner arter çıkışı anomalisi; koroner arter fistülü; ekokardiyografi.
Coronary artery anomalies are a diverse group of congenital disorders, in terms of morphological characteristics, pathophysiological mechanisms, diagnostic work-up, prognosis, and treatment requirements.[1] These anomalies are reported in 0.6 to 1.5% of the coronary angiograms in the literature.[2] They can be classified into two main groups as anomalies of origin and distribution and as anomalies of inter-coronary communications and coronary artery fistulas.[3] Coronary arteriovenous fistulas (CAFs) are abnormal connections of coronary arteries with cardiac chambers or vascular structures.[3] The incidence is around 1/50,000 at birth and 1/500 at cardiac catheterization, 0.3 to 0.4% in patients with congenital heart defects.[4,5] In one study from Turkey, the angiographic incidence and rate of CAFs was found to be 0.04% and the rate of CAFs was 5% among coronary artery anomalies.[2,5] The angiographic incidence and percentage of CAF among coronary artery anomalies were found to be higher in another study from Turkey (0.45% and 18.6%, respectively).[5,6]

Coronary arteriovenous fistulas cause blood flow to be shunted into cardiac chambers, great vessels or other structures, passing the myocardial capillary network. Symptoms may be caused by either coronary steal or a high left-to-right shunt flow.[1] The majority of the cases are asymptomatic; however, with exercise, symptoms such as chest pain, dyspnea, and syncope can be manifested.[7-10] In CAFs, clinical symptoms and the age of onset are associated with the underlying anatomy and the diameter of the fistula.[8] When calibration of the fistula is small, clinical findings may not be evident. If the diameter of the fistula is large, mitral and tricuspid regurgitation, heart failure, myocardial ischemia, infarct, bacterial endocarditis, rupture and premature arteriosclerosis may be present.[3,8] The aforementioned clinical findings and complications can be avoided with early closure of the fistula. In addition, fistulas may originate from the right coronary artery (RCA) (50%), left main coronary artery (LMCA) (42%) or both (5%) based on their frequencies. They usually drain into the right side of the heart (92%), followed by the right ventricle (RV) (41%), right atrium (RA) (26%), coronary sinus (CS) (7%), pulmonary artery (PA) (17%), and superior vena cava (SVC) (1%) respectively. They rarely drain into the left cardiac chambers (8%).[3]

Anomalous origin of left coronary from pulmonary artery (ALCAPA) and anomalous origin of right coronary artery from pulmonary artery (ARCAPA) can be grouped in anomalous origination of coronary artery. The former is anomalous origin of LMCA arising from PA and the latter is anomalous origin of RCA arising from PA.[11-13] The estimated incidence of ALCAPA is 1/300,000 live births.[14] In addition, ARCAPA is rare than ALCAPA. Clinical findings can be similar to those in large CAFs. Myocardial ischemia and infarct, congestive heart failure, cardiogenic shock, and sudden death may be seen in ARCAPA. Conversely, heart failure and ischemia development are not expected in ARCAPA and sudden death rate is relatively low.[13]

Origination of LMCA from the right sinus of Valsalva with a route between PA and aorta is another anomaly of origination from aorta leading to ischemia and sudden death with compression between these vessels establishing with an incidence of 0.03 to 0.12%.[4,15]

In the present study, we aimed to evaluate patients with CAFs, ALCAPA, and ARCAPA and to investigate the role of echocardiography (ECHO) in the diagnosis.

**PATIENTS AND METHODS**

**Clinical data collection**

Fourteen cases who were referred to pediatric cardiology clinic with CAFs, ALCAPA, and ARCAPA between January 2009 and March 2015 were retrospectively analyzed. Patients with coexistent congenital heart diseases were excluded. The patients were evaluated based on their clinical findings, symptoms, and physical examination findings which were supported with electrocardiography (ECG), telecardiography (TELE), transthoracic echocardiography (TTE), conventional angiography, and cardiac computed tomography angiography (CTA), when necessary. The study was conducted in accordance with the principles of the Declaration of Helsinki.

All patients were divided into two groups as those with CAFs (group 1) and those with anomalous origin of coronary artery from pulmonary artery (group 2). Group 1 consisted of 10 (5 males, 5 females; mean age 10.3±8.5 years; range 1.5 to 32 years) and group 2 consisted of four (1 male, 3 females; mean age 9.5±5 years; range 3 to 15 years) patients. Clinical characteristics of the patients, diagnostic work-ups, and treatment decisions are summarized in Tables 1 and 2.

According to the complaints, the patients in group 1 were consulted for chest pain with and without exercise, murmur, and palpitation. The patients in group 2 were consulted for chest pain and syncope during exercise. One other patient in group 2 was referred for dilated cardiomyopathy (DCMP).
<table>
<thead>
<tr>
<th>Connection</th>
<th>Patient no</th>
<th>Gender</th>
<th>Age (year)</th>
<th>Symptoms/Physical examination</th>
<th>Trans-thoracic echocardiography</th>
<th>Computed tomography angiography</th>
<th>Qp/Qs Angiography</th>
<th>Decision</th>
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<tbody>
<tr>
<td>LAD-PA</td>
<td>1</td>
<td>M</td>
<td>8</td>
<td>CP/N</td>
<td>Cardiac chambers-upper border, Diastolic flows in RV+PA, Dilated LMCA</td>
<td>-</td>
<td>1.2</td>
<td>Clinic follow-up</td>
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<td></td>
<td>4</td>
<td>F</td>
<td>7</td>
<td>CP/N</td>
<td>Cardiac chambers-N, Continuous flows → PA more visible in diastole</td>
<td>-</td>
<td>1.1</td>
<td>Clinic follow-up</td>
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<td>5</td>
<td>F</td>
<td>7</td>
<td>RC/N</td>
<td>Cardiac chambers-N, Continuous flows → PA more visible in diastole</td>
<td>-</td>
<td>1.1</td>
<td>Clinic follow-up</td>
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<tr>
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<td>2</td>
<td>M</td>
<td>8</td>
<td>CP/M</td>
<td>Cardiac chambers-N, Continuous flows → PA more visible in diastole</td>
<td>-</td>
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<td>Clinic follow-up</td>
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<td>3</td>
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<td>12</td>
<td>CPWE/M</td>
<td>Cardiac chambers-N, Continuous flows → PA more visible in diastole</td>
<td>-</td>
<td>1</td>
<td>Clinic follow-up</td>
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<td></td>
<td>10</td>
<td>M</td>
<td>13</td>
<td>P/M</td>
<td>Cardiac chambers — Slight dilatation, Continuous baffle leak flow intrapulmonary tunnel (7 mm)—PA, MR (1st degree)</td>
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<td>1.5/Proximal LMCA dilatation (secondary due to operation)</td>
<td>Clinic follow-up (Surgery was refused)</td>
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<tr>
<td>RCA-RV</td>
<td>6</td>
<td>F</td>
<td>1.5</td>
<td>M/M</td>
<td>Left chambers-Dilated, Dilated RCA (11 mm-wp, Continuous flow in RA (2.8 mm-mp)</td>
<td>Dilated vascular structure (12.5 mm) RCA-RA</td>
<td>2.5/Aneurysmatic dilatation of RCA (6.25 mm-wp /3.76 mm-mp)</td>
<td>Embolized- 5x6 ADO-II</td>
</tr>
<tr>
<td></td>
<td>7</td>
<td>F</td>
<td>11</td>
<td>M/M</td>
<td>Right cardiac chambers—upper border, Ectasic RCA origin, Continuous flows on RCA</td>
<td>-</td>
<td>1.5/Aneurysmatic dilatation of RCA (7-8 mm-wp)</td>
<td>Embolized- 5x5 mm Cook Flipper coil</td>
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<td>RCA-CS</td>
<td>8</td>
<td>M</td>
<td>3</td>
<td>M/M</td>
<td>Cardiac chambers-N, Ectasic RCA, Continuous flow in RV</td>
<td>-</td>
<td>2/Ectasic RCA, Absent PDA</td>
<td>Embolized- 5x5 mm Cook Flipper coil</td>
</tr>
<tr>
<td>RCA-CS</td>
<td>9</td>
<td>M</td>
<td>32</td>
<td>M/M</td>
<td>Cardiac chambers-dilated, Very dilated CS (28 mm), Continuous flow draining to CS and RA, Aneurysmatic dilatation on proximal of RCA (12 mm-at the begining)/20 mm-dubuent dilatation</td>
<td>Aneurysmatic RCA opening to enlarged CS, Very dilated RCA, Aneurysmatic sac like formation associated with CS</td>
<td>1.7/Very enlarged RCA origin (18 mm), Abnormal trace of RCA, Tortuous RCA, Sac like shape of RCA before opening to dilated CS (0 mm), Stenosis at the opening of RCA to CS</td>
<td>Pure-string ligation-for the orific of CAF, Plication for the aneurysmatic segment of RCA</td>
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CTA: Computed tomography angiography; Qp/Qs: Pulmonary/systemic flow rate; LAD: Left anterior descending; w/p: Widest part; PA: Pulmonary artery; CP: Chest pain; N: Normal; RV: Right ventricle; LMCA: Left main coronary artery; RC: Routine control; M: Murmur; CPWE: Chest pain with exercise; MR: Mitral regurgitation; RCA: Right coronary artery; RA: Right atrium; n-p: Narrowest point; ADO: Amplatz duct occluder; CS: Coronary sinus; PDA: Posterior descending artery; CAF: Coronary arteriovenous fistula.
The patients in group 1 who had intervention and all patients in group 2 were diagnosed with the help of a typical auscultatory finding of a systolic-diastolic murmur over precordium. Two patients (Cases 2 and 3) had systolic murmur, while the remaining three patients did not have any auscultatory finding.

Electrocardiography findings were non-specific in the patients of group 1. One patient had incomplete right bundle branch block in leads V1 and V2 in group 1. Patients, TEL, showed no significant findings, while it showed dilated cardiac silhouette in one patient in group 2. In addition, cardiac CTA was performed in two patients in group 1. It revealed fistulas between RCA and RV (Figures 1 and 2) and RCA and CS (Figure 3a). Before opening to dilated CS, RCA was extremely dilated. Two-dimensional (2D) ECHO studies, Doppler sonography, and color Doppler ECHO were used in the patients of group 1. Three patients had absent LMCA origin from aorta, dilated RCA, collateral arteries filling LMCA, LMCA flows draining to PA, and LMCA originating from PA. Absent LMCA origin from aorta, dilated RCA, collateral arteries filling LMCA, LMCA flows draining to PA, and LMCA originating from PA. LMCA was reimplemented to aorta. Absent LMCA origin from aorta, dilated RCA, collateral arteries filling LMCA, LMCA flows draining to PA, and LMCA originating from PA. LMCA was reimplemented to aorta.
dilated and tortuous with a small aneurysmal sac-like formation (Figure 3b-d). Only one patient in group 2 underwent CTA which showed ecstasia in RCA with absent LMCA origin from the aorta. Cardiac catheterization and angiography was performed to all patients. Oxymetrically calculated shunt volumes from the left to right defined as $Q_p/Q_s$ were obtained. The $Q_p/Q_s$ was ≥1.5 in five patients in

![Figure 2](image1.png)

**Figure 2.** Absent flow in the right coronary artery after embolization the fistula between right coronary artery and right ventricle (Case 8).

![Figure 3](image2.png)

**Figure 3.** (a) An echocardiographic image showing the right coronary artery fistula opening to coronary sinus. (b) Dilated RCA and dilated CS are shown in three-dimensional CTA. (c) Dilated and tortuous RCA in the CTA, an aneurysmal sac-like formation associated with dilated CS. (d) Angiographic image showing aneurysmal and dilated RCA (Case 9).

RCA: Right coronary artery; CS: Coronary sinus; CTA: Computed tomography angiography.
group 1. According to the angiographic findings, four patients (40%) had a fistula originating from the RCA; three (30%) from the left anterior descending (LAD) artery; two (20%) from the LMCA, and one (10%) from the ‘baffle’ between PA and proximal part of the LMCA. Fistulas drained into the RA in two (20%), RV in one (10%), CS in one (10%), and PA in six patients (60%).

Aortic root and selective coronary artery angiograms showed a dilated RCA arising from the aortic root without normal origin of LMCA which was filled later via coronary artery collaterals and ultimately shunted into the PA in the patients with ALCAPA.

**Treatment**

The shunt volume was low (Qp/Qs <1.5) in five patients and symptoms were not associated with cardiac causes; therefore, they were followed in the outpatient setting and no intervention was performed. The remaining five patients had Qp/Qs ≥1.5. Three of these patients underwent transcatheter approach, while one patient underwent surgery. One patient who was indicated for surgery did not accept treatment and was followed in the outpatient setting. A Flipper coil (Cook Medical, Bloomington, IN) was used in two patients and an Amplatzer Duct Occluder II (ADO) (AGA Medical Corporation, Plymouth, MN, USA) device was used in another. A 5x6 ADO-II was used in the patient who had a fistula with diameters as 6.25 mm at the widest and 3.76 mm at the narrowest part. A Cook Flipper coil 5x5 mm was used in two patients. The orifice of the fistula was measured as 3.8 mm at the opening point to the RV in one patient (Case 8). The fistula was 8 mm at the widest part in another patient (Case 7). Every fistula was occluded by only one occluder. The ADO-II was preferred for larger and Cook coil for smaller fistulas. Surgery was applied due to the tortuous anatomy of the fistula in a patient (Case 9). Porse-string ligation was performed for the orifice of the fistula between the RCA and CS under cardiopulmonary bypass (CPB) and cardioplegic arrest. Plication was applied for the aneurysmatic segment of the RCA. Surgical treatment was also indicated for the patient who had Takeuchi operation for ALCAPA at eight years old with a present baffle leak between the PA and proximal part of the LMCA. However, this patient refused treatment.

In addition, three patients with ALCAPA and one with ARCAPA were treated surgically. Surgery was also performed under CPB and cardioplegic arrest. The PA was transected above the level of the pulmonary valve. The left or right coronary artery orifice was resected from the PA with a button of surrounding PA tissue and, then, using end-to-side anastomosis with a punched out opening in the left posterolateral wall or anterior wall of the ascending aorta. The defect in the PA was repaired with a generous fresh autologous pericardial patch.

**RESULTS**

All patients who underwent treatment procedures survived. Three patients with transcatheter approach had a successful occlusion. All of them demonstrated complete fistulas closure. No peri-procedural death events occurred. Technical success was achieved in all three patients. The closure was performed applying a coil and ADO, and acetylsalicylic acid was administered.

The patients were discharged on the next day. One patient who underwent surgery for a CAF was also discharged without any complication after one week. There were no in-hospital mortality and no major complications in the patients with ALCAPA and ARCAPA. None of these patients required extracorporeal membrane oxygenation after the operation. They were all discharged within the postoperative fifth and eighth days.

The mean follow-up was 4.4±1.8 (range, 2 to 6 years) following the interventional procedures in group 1. All of the patients with transcatheter occlusion in group 1 were followed in the outpatient setting and using ECG, TELE, ECHO at one, three, six and 12 months during the first year and annually, thereafter. All patients were asymptomatic with no clinically audible murmurs. On follow-up ECHO, no recanalization of the fistula, coil migration or occluder displacement or fractures were shown. The patient with a fistula closed by ADO-II also received warfarin, as assessed by repeated angiography for the ongoing fusiform dilatation on the RCA proximal. However, as the patient was lost to follow-up, disease progression is not known. One patient who did not accept surgical treatment was under medical follow-up for MR and dilatation of the left cardiac chambers. No progression was seen in the clinical findings. Initial complaints showed regression in the patients who were followed in the outpatient setting.

The mean follow-up was 2.7±2.4 (range, 0.5 to 6 years) following surgical procedures in group 2. The patient with ARCAPA and those with ALCAPA, except one (Case 13) attended their scheduled follow-up visits regularly. Cardiac functions were affected only in one patient (Case 13) with left ventricular ejection fraction...
(LVEF) of 60% and fractional shortening (FS) of 30% with improved outcomes two weeks after the operation (LVEF: 62%, FS: 32%); however, the patient was lost to follow-up. The size of cardiac chambers returned to normal limits in the remaining patients with reduction in MR and there were no ischemia and arrhythmia findings.

**DISCUSSION**

Coronary artery anomalies should be regarded as an uneven diverse group of congenital disorders of which manifestations and pathophysiological mechanisms are highly variable.[1] Although coronary artery anomalies are often asymptomatic, some may be evident with clinical findings or sudden death. Therefore, the recognition of coronary anatomical patterns is of utmost importance, particularly when identifying patients who need intervention or not.[5]

Two dimensional TTE indicates the suspicion of CAF, when dilated coronary arteries are accompanied with symptoms and clinical findings. The entire epicardial trace and end point of the fistula, and continuous or turbulent flow within the fistula or at the site of termination can be seen with pulsed Doppler and color Doppler ECHO. If shunt volume, pressure, and systemic and pulmonary resistances. Therefore, aortography and selective coronary angiography are required for typing the CAF defect and planning the treatment approach.[9] It can be used for therapeutic embolization with occlusive coils and devices, as well. In certain cases, the origin and relation of the CAF to the adjacent structures may be difficult to determine; therefore, non-invasive techniques may be used as adjunct.[16] Also, it provides a two-dimensional representation of coronary course.[15]

Three-dimensional (3D) construction of the anatomy with improved delineation and spatial resolution can be obtained with CTA, compared to coronary angiography.[17]

Accordingly, in our patients with fistula, the diagnosis was suspected upon stating enlarged cardiac chambers and continuous flows in cardiac chambers, PA and CS with TTE. Then, CTA was performed in two patients and conventional angiography in all patients in group 1. The former was performed due to a suspicion with TTE to confirm the diagnosis and to have a better orientation for the anatomy of the related coronary artery and fistula with a 3D definition to tailor the treatment approach. In our study, catheter angiography was applied to the patients, as it was considered as the gold standard approach for fistulas and to clarify their symptoms which were unable to be explained with other reasons, to make a definite diagnosis, to assess other additional anomalies, and to make hemodynamic evaluation by calculating the shunt volume, pressure, and resistance. Consequently, CAFs originated from the LAD and LMCA in 50% and from the RCA in 40% of our patients, whereas they usually originate from the RCA as reported in the literature.[18] The difference can be explained by the limited number of the patients in the study. In addition, CAFs drained into the right side of the heart, supporting the literature data.

Furthermore, as pulmonary vascular resistance decreases after birth, the retrograde flow through the anomalous coronary artery from the collateral circulation mostly floods inward into the PA, resulting in coronary steal and subsequent angina.[18] Compared to ALCAPA, ARCAPA less often results in the classical syndrome of myocardial ischemia, infarction, and sudden death.[9] Some authors have hypothesized that this is due to the lower oxygen demand of the right ventricle, compared to the left ventricle and the smaller amount of myocardial territory fed by the RCA compared to the LMCA.[13] The suspicion is made when the origins of coronary arteries in patients with ALCAPA and ARCAPA cannot be visualized with TTE. Secondary ECHO findings such as MR, ventricular dilatation, coronary collaterals within IVS (Figure 4), dilatation of the other coronary artery (Figure 5), originating of the anomalous coronary artery from PA (Figure 6), subendocardial fibroelastosis, and papillary muscle dysfunction support the diagnosis. A flow toward the probe can be visualized at the PA above pulmonary valve. If ALCAPA is highly suspicious or diagnosed based on ECHO findings,
it should be confirmed by conventional angiography before cardiac surgery. Left-to-right shunting blood flow can be seen, PA pressure can be obtained, and abnormal origin of LMCA (Figure 7), dilated RCA (Figure 8) and collateral flows can be detected with angiography. Of note, CTA is an alternative imaging technique that is used for diagnosis. It is valuable in visualizing abnormal coronary arteries, their origins, and projections. Also, the sensitivity of angiography may be limited in the diagnosis of an anomalous coronary artery due to its invasive nature. Therefore, CTA and/or magnetic resonance imaging is recommended for more definite spatial resolution before deciding surgery. With the recent developments, ECG-gated multi-detector computed tomography angiography (MDCT) also provides non-invasive diagnosis in coronary artery diseases and malformations. It is believed that non-invasive techniques can replace conventional angiography for the definitive diagnosis.

All of the patients in group 2 showed the diagnostic findings with TTE. None of the patients were diagnosed with ALCAPA or ARCAPA initially with CTA without any suspicion on TTE. In one patient with ALCAPA, CTA was performed to have a clear visualization of the coronary artery before surgery. All patients in group 2 underwent conventional angiography to make the definite diagnosis.
The American Cardiology College/American Heart Association (ACC/AHA) (2008) recommends transcatheter or surgical closure for all large coronary artery fistulas independent from the presenting symptoms. Small and medium-sized fistulas are recommended to be closed only in the presence of symptoms (i.e., ischemia, arrhythmia, unexplained systolic and diastolic dysfunction). Our patients who required intervention were symptomatic and had a prominent left-to-right shunt as confirmed by angiography with ≥1.5 Qp/Qs in all patients.

Surgical ligation of fistulas is one of the treatment methods. Although surgery is safe and effective with good results, transcatheter occlusion of the fistula in pediatric patients is recommended as the preferred treatment. Compared to surgery, transcatheter closure is less invasive, less prone to complications, and has a shorter hospital stay. Therefore, transcatheter closure is an emerging alternative in patients, when the anatomy of the fistula is favorable for this treatment (e.g., non-tortuous vessel, the fistula should be unique with distal narrowing to avoid embolism to the drainage site, distal portion of the fistula should be accessible with the closure device, and there should be no concomitant cardiac disorders requiring additional surgical intervention). Coils were used in the majority of patients in the literature. Vascular plugs, umbrella devices, and covered stents are also used. The main limitations of the transcatheter approach, however, are inability to deliver the catheter to the fistula due to excessive tortuosity, inadequate size of the coronary artery, and presence of coronary branches at the site of the CAF, which occur in nearly 15% of cases.

The Amplatzer PDA occluders have several advantages over other devices to close CAFs, including the use of a single device, a high rate of complete occlusion, and improved control over placement and release of the device. In addition, these devices are used for saccular or aneurysms before the fistulous orifice.

Armsby et al. reported short-term results in 33 patients who underwent transcatheter closure which showed no flow in 22 of them and small residual shunts in five. None of these patients attended to the follow-up angiography. Complete closure was 91% during a mean time of 12 months. In our study, coils were used for small fistulas, whereas ADO devices were used for larger ones. The fistula that ADO was used for closure had an accompanying aneurysm, similar to the literature reports.

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Patients who underwent interventional procedure used acetylsalicylic acid at anti-thrombotic doses (3 to 5 mg/kg/day) to prevent thrombosis. The potential causes of the mechanism of thrombus formation after closure of the CAFs include the reduction of blood flow in the aneurysmal artery. Certainly, stasis of flow can occur with long-standing CAF and consideration should be, therefore, given to oral anticoagulation. The duration of antiplatelet or anticoagulant therapy following the procedure is unclear; however, it is reasonable to continue at least four week and perhaps longer. The patient who received ADO used both warfarin and acetylsalicylic acid for the ongoing dilatation of the coronary artery.

Current treatment options for CAFs include surgical ligation (either with or without CPB), and surgical ligation accompanied by coronary artery bypass grafting. In most cases CPB is needed to open the cardiac chamber to find and ligate the points of entry and to avoid postoperative ischemia and arrhythmia. In the present study, one patient was treated surgically due to the aneurysmatic and tortuous structure of the CAF under CPB with surgical ligation alone without any postoperative problem. The patient with fistula between the LMCA proximal and PA consisting of a ‘baffle’ leak secondary to Takeuchi operation refused
surgical treatment. Transcatheter procedure was not considered for treatment, as the origin of the baffle leak was very close to the LMCA orifice and a device to be used to terminate the flow could potentially narrow the LMCA, thereby, leading to ischemic findings.

Furthermore, surgical treatment is recommended due to cardiac symptoms and sudden death risk in ALCAPA and ARCAPA, which are anomalies of origin of coronary arteries. Patients with ALCAPA should be operated at the time of diagnosis, as the mortality rates are very high, although they are asymptomatic. Most of the affected patients show symptoms in infancy and early childhood. About 90% of untreated infants die within the first year of life and only a few patients survive until adulthood. On the other hand, although ARCAPA was considered historically as a benign lesion, it was associated with cardiac symptoms and sudden death. The improvements in cardiothoracic surgery, cardiac anesthesiology, and intensive care units have reduced the morbidity and mortality rates of ARCAPA surgery. Therefore, according to the risk-benefit ratio, surgery is recommended for ARCAPA. Surgery can be planned, when the infants reach a size, where the risks of coronary reimplantation are minimized. They can be treated medically until operation. Therefore, surgery can be delayed after infancy period, if they remain asymptomatic. Successful surgical repair basically depends upon the establishment of a dual coronary artery system. The surgical procedure may eliminate the steal syndrome and restore physiological antegrade flow. It can also restore and maintain the LVEF, decrease ventricular dilatation, relieve angina, protect the mitral apparatus, prolong life expectancy, and achieve good early and late results. There is a controversy as to whether or how, the mitral valve is to be repaired at the time of coronary reimplantation. Some authors recommend routine mitral valve repair at the time of coronary artery revascularization, while some advocate mitral annuloplasty or mitral valve replacement only in the presence of severe MR. Some others do not recommend repairing moderate or severe MR at the initial ALCAPA repair. In pediatric patients, even severe MR was reported to fully regress after reperfusion alone in the majority of the cases. Our opinion is that the patients can be better candidates for a definitive mitral valve repair or replacement, if they are older, they have recovered from the myocardial ischemia caused by ALCAPA, and their left ventricles have improved or returned to normal function particularly in the infancy period.

The patient with ARCAPA was an adolescent at time of diagnosis and underwent surgery. On the other hand, the patients with ALCAPA were operated soon after the diagnosis. None of them needed mitral valve surgery, as they had mild or moderate MR. Moderate MR regressed to mild six months after surgery. The diameters of the cardiac chambers were observed to decrease in the patients who attended to scheduled follow-up visits. Cardiac functions were better in the patient with lower LVEF two weeks after the operation.

In conclusion, although coronary artery fistulas are usually asymptomatic without any need for treatment, they should be considered in cases particularly with enlarged cardiac chambers and dilated coronary arteries, since they can be easily overlooked due to their low incidence. A careful evaluation should be made with transthoracic echocardiography. Cardiac computed tomography angiography is an alternative imaging option in children. Cardiac catheterization and angiography should be considered and treatment options should be applied in selected cases. The choice of device and technique for any patient is based on many factors, particularly including the anatomic characteristics of the fistula. In our patients, transcatheter closure using either the Cook coil or Amplatzer duct occluder is suggested to be a safe and effective method. Based on our clinical experience, transcatheter closure of coronary artery fistulas with either coils or occluders is a feasible alternative to surgery with excellent short-term outcomes. Surgery should be performed in selected patients. Anomalous origin of the left coronary from pulmonary artery and anomalous origin of the right coronary artery from pulmonary artery should be treated using an arterial reimplantation technique preferably, to create a dual coronary arterial system based on the individual anatomical structures. According to these findings, we conclude that echocardiography still has the main role in identifying these rare, but critical life-threatening diseases.

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