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Coronary-Pulmonary Artery Fistulas In Children: A Single-Center Experience

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ABSTRACT

This study aims to evaluate the characteristics and outcomes of children with coronary to pulmonary artery fistulas.
The age, gender, duration of follow-up, indications for initial echocardiography, electrocardiography and echocardiography findings, catheter angiography indications, and the outcome of patients were evaluated. Echocardiographic data included additional cardiac anomalies, origin and drainage of the fistulas, presence of coronary artery dilatation, and changes in fistula flow.
The study involved 64 (66.7%) male and 32 (33.3%) female children. Additional cardiac anomalies were present in 34 (35.4%) cases. The origin of the fistula was defined in only 9 cases (by echocardiography and angiography) and the site of origin was: the left anterior descending (55%), the left circumflex (2.5%), and the right coronary (2.5%) artery. Two cases had very mild dilatation of coronary arteries. The drainage of the fistulas was to the anterior aspect of the pulmonary artery (78.7%), the aortic side of the pulmonary artery (4.3%), and the right pulmonary artery (17%). In only 2 cases, catheter angiography was performed with an indication (suspected coronary artery anomaly) related to coronary-pulmonary fistula, and one of them had right coronary artery dilatation. Rate of spontaneous closure was 9.4%. Surgical or percutaneous closure was not considered in any of the cases and none of the cases had any adverse events or symptoms related to the fistula.
Coronary to pulmonary artery fistulas seldomly require further investigation. Clinical follow-up with echocardiography and ensuring bacterial endocarditis prophylaxis is usually adequate unless the fistulas are hemodynamically significant and/or symptomatic.

Keywords: Coronary artery; coronary-pulmonary fistula; coronary dilatation; cardiac anomaly

Introduction

Coronary artery fistula (CAF) is an abnormal connection between the coronary artery and a cardiac chamber or systemic/pulmonary circulation. The incidence of CAF in children undergoing echocardiography is reported as 0.06% (1, 2). Although most fistulas are small in size and asymptomatic, larger fistulas may be hemodynamically significant and cause symptoms due to coronary steal phenomenon and/or volume overload (2). The complication rate of the fistulas is 11% under the age of 20 years, whereas it increases to 35% above the age of 20 years (3). Though it is rare, infective endocarditis and endarteritis should be kept in mind as important complications (2, 4). Although surgical ligation or percutaneous closure of large and symptomatic fistulas is advised, there is no consensus on the treatment of incidentally detected small fistulas (3, 5).

Approximately 90% of CAF are congenital, and 20% are associated with other cardiac anomalies. Acquired fistulas may be secondary to various conditions such as coronary intervention, radiation, infective endocarditis, pacemaker implantation, vasculitis, myocardial infarction, cardiac surgery, and Kawasaki disease (1-5).

Coronary-pulmonary artery fistula (CPAF) is an abnormal connection between the coronary arteries (CA) and pulmonary arteries (PA) and accounts for 15-30% of all CAF (2, 6). It is the most common type of CAF observed in children and its prevalence is estimated as 0.17%-0.68% (4). They are mostly small, and diagnosis is made incidentally during the echocardiographic evaluation of the patient for other reasons (1-5). The incidence of CPAF has increased over decades with the advent of imaging modalities but their clinical significance is still unclear.

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In this retrospective study, we aimed to evaluate the characteristics and outcomes of children with CPAF.

Material and Methods

Medical records of all patients that were diagnosed and followed up with CPAF at the Pediatric Cardiology Department of our institution between 1993-2020 were reviewed retrospectively. The local ethics committee approved the study. (16.02.2022 / İ02-93-22)

All patients with diagnosis of fistula in our database were included in the study except the patients with fistula other than CPAF, with a follow-up period of less than one year, and with missing data.

The age, gender, height, and weight at the time of diagnosis, duration of follow-up, indications for initial echocardiography, electrocardiography and echocardiography findings, catheter angiography indications, and the outcome of the patients were evaluated.

All of the cases were evaluated by electrocardiography and echocardiography. Echocardiographic evaluation of all cases was performed according to the guidelines of the American Society of Echocardiography (7). Echocardiographic data included origin and drainage of the CPAF, presence of coronary artery dilatation, changes in fistula flow, and additional cardiac anomalies that were present at the time of admission or diagnosed during the follow-up period. Small atrial septal defects (ASD), hemodynamically insignificant very small patent ductus arteriosus (PDA), and very small ventricular septal defects (VSD) were not included as additional cardiac anomalies.

Statistical analyses were performed using the statistical package program SPSS for Windows 22.0 (IBM Corp., Armonk, NY, USA). The mean, standard deviation, and frequency were used for descriptive statistics.

Results

There was a total of 129 cases diagnosed with fistula in our database. Thirty-three cases with insufficient data and/or fistulas other than CPAF were excluded from the study.

As a result, the study included 64 (66.7%) male and 32 (33.3%) female cases. The mean age at the time of diagnosis of CPAF was 66.62 ± 58.36 months. Mean height and weight at the time of diagnosis were 97.87 ± 36.99 cm and 18.91 ± 16.14 kgs, respectively. The mean duration of follow-up after the diagnosis was 35.53 ± 34.54 months (range:12 to 187 months).

The indications for initial echocardiographic evaluation are shown in Table 1. None of the cases had any symptoms and/or electrocardiography findings that were attributed to the presence of CPAF. Additional cardiac anomalies were detected in 34 (35.4%) cases at the time of diagnosis and/or during the follow-up period. The distribution of additional cardiac anomalies is shown in Table 2. Five cases with cardiac anomalies that required surgical management were as follows: Atrioventricular septal defect (n:1), VSD (n:1), ASD (n:1), PDA (n:2). Percutaneous PDA closure was performed in 5 cases. Spontaneous closure of VSD was observed in two cases. One case with dilated cardiomyopathy died during follow-up. Four cases were diagnosed as Down syndrome, whereas one case was diagnosed as Beckwith-Wiedemann syndrome.

Sixty-nine (71.9%) of the cases in our study were diagnosed at the initial echocardiographic examination and 92.9% were diagnosed at the first five echocardiographic examinations. All the cases that were not diagnosed at the initial evaluation also had additional cardiac anomalies and some of the cases were diagnosed after surgical, percutaneous, or spontaneous closure of left to right shunt defect.

The origin of CPAF was defined only in 9 cases by echocardiography and catheter angiography: 5 (55%) were originating from the left anterior descending artery (LAD), 2 (22.5%) from the left circumflex artery (LCx), and 2 (2.5%) from the right coronary artery (RCA). Only two cases had very mild dilatation of coronary arteries. The case with dilatation of RCA was evaluated with catheter angiography and the other case with dilatation of LAD was evaluated with computed tomography angiography (CTA).

Catheter angiography was performed in a total of 12 cases. In 10 of these cases, the indication for catheter angiography was congenital heart disease (CHD) but in the remaining 2 cases catheter angiography was performed with an indication related to CPAF. In these two cases, catheter angiography was performed because of suspected coronary artery anomaly and one of these cases had RCA dilatation.

Spontaneous closure of CPAF was observed in 9 (9.4%) cases and 5 of these cases also had

Table 1. Initial Echocardiography Indications

	n (%)
Murmur	48 (50%)
Congenital heart disease	12 (12.5%)
Syndromic appearance	7 (7.3%)
Newborn screening	6 (6.3%)
Chest pain	6 (6.3%)
Chemotherapy side effect	4 (4.1%)
Suspected acute rheumatic fever	2 (2.1%)
Etiology of fever	2 (2.1%)
Shortness of breath	2 (2.1%)
Palpitation	2 (2.1%)
Fetal arrhythmia	1 (1.04%)
Short stature	1 (1.04%)
Hypertension end organ damage	1 (1.04%)
Elevation of cardiac enzymes	1 (1.04%)
Routine cardiac control	1 (1.04%)

Table 2. Distribution of Associated Cardiac Anomalies

	n (%)
Atrial septal defect	9 (9.3%)
Patent ductus arteriosus	7 (7.3%)
Ventricular septal defect	7 (7.3%)
Mitral valve prolapse	6 (6.3%)
Bicuspid aortic valve	5 (5.2%)
Dilatation of sinus Valsalva	2 (2.1%)
Rheumatic heart disease	1 (1.04%)
Dilated cardiomyopathy	1(1.04%)
Non-compaction cardiomyopathy	1 (1.04%)
Atrioventricular septal defect	1 (1.04%)
Subaortic stenosis	1 (1.04%)
Right aortic arch	1 (1.04%)

additional cardiac defects: VSD (n:1), ASD (n:3), and Mitral valve prolapse (n:1).

Discussion

Coronary to pulmonary fistula is a rare anomaly of the coronary arteries. Several studies have reported different prevalence rates due to the variations in the diagnostic tools used (8-10). Most patients with CPAF are asymptomatic and are mostly diagnosed incidentally. Thus, it is difficult to estimate the true incidence of CPAF, although the incidence has increased over the decades with the advent of imaging modalities. Echocardiography is the first step non-invasive imaging modality that is routinely used in the diagnosis and follow-up of cardiac diseases. In our study, 59 (61%) of the cases diagnosed with CPAF underwent echocardiography because of symptoms such as murmur, chest pain, and palpitation. In the rest of the patients, fistulas were detected during evaluation for other cardiac diseases. Considering that none of the patients had any symptom that was attributed to the presence of fistula, we think that all the cases included in our study were diagnosed incidentally.

No race or sex predilection has been reported for CAF (3). However, there was a male predominance (66.7%) in our study and Ugan-Atik et al. (11) reported a similar rate (63.3%) of male predominance in their study evaluating children with CPAF.

Coronary to pulmonary fistulas may be associated with other cardiac anomalies, and Wong et al. (12) and Schumacher et al. (13) reported the rate of an additional cardiac anomaly in children with CPAF as 38% and 27%, respectively. In our study, this rate was 35.4% and the most common anomalies were ASD, VSD, PDA, mitral valve prolapse, and bicuspid aortic valve. Similarly, Ugan-Atik et al. (11) reported VSD, PDA, and ASD as the most common additional cardiac anomalies in children with CPAF. This result is not surprising because CPAF is mostly an incidental finding, and VSD, ASD, and PDA are also the most common cardiac anomalies in children.

The clinical manifestations depend on the amount of left to right shunt and/or the extent of the coronary steal which are both determined by the size of the CPAF. It may cause murmur, palpitation, chest pain, easy fatigue, failure to thrive, and shortness of breath (2, 4). However, CPAF is usually an incidental finding and at least 75% of patients are asymptomatic (10). In our study, murmur (50%), chest pain (6.3%), palpitation (2.1%), and shortness of breath (2.1%) were among the indications for the initial echocardiographic evaluation, but none of these complaints were attributed to the presence of CPAF.

Sixty-nine (71.9%) of the cases in our study were diagnosed at the initial echocardiographic examination and 92.9% were diagnosed at the first five echocardiographic examinations. Similarly, Ugan-Atik et al. (11) reported that 78.2% of the cases in their study were diagnosed at the initial examination and the latest diagnosis was made at the 10th examination. Besides the fact that these fistulas are usually seen as very small flows into the PA; the late diagnosis can result from focusing mostly on the main cardiac anomaly, which may have resulted in the failure to notice these small fistulas. The presence and the retrograde flow of PDA into the pulmonary artery, turbulent flow in the PA, and elevated PA pressure due to significant left to right shunt may also mask the flow of the fistula. Here, one can also question if these CPAF were acquired or congenital.

It is not always possible to define the coronary origin of CPAF with echocardiography because they are usually seen as very small flows into the pulmonary artery. Liu et al. (6) evaluated 43 adult patients with CPAF with coronary angiography and reported that LAD (41.67%) was the most common site of origin followed by the RCA (38.89%), the left main coronary artery (12.5%), and the LCx (6.94%). A recent systemic review reported the left main coronary artery (84%) and the RCA (38%) as the most common origins of CPAF (14). Although we were able to define the coronary origin of fistula with echocardiography and angiography only in a few numbers of cases, LAD (55%) was the most involved coronary artery followed by LCx (22.5%) and RCA (22.5%).

The main pulmonary trunk is the most common site of drainage of CPAF (2, 15). Hang et al. (16) reported the most common drainage site of CPAF as the left anterior PA (86.36%), the anterior wall of PA (9.09%), and the left PA (4.55%). Ugan-Atik et al. (11) reported that the fistulas drained to the anterior aspect of PA (69.3%), the aortic side of PA (22.8%), and the right PA (7.9%). When classified in the same way, these ratios were 78.7%, 4.3%, and 17% respectively in our study but none of the fistulas were draining to the left PA. As previously reported in the literature, the main pulmonary trunk was the most common site of drainage in our study, however drainage of fistulas into the right PA was more common than the aortic side of main PA.

The rate of coronary dilatation was 2.1% in our study, however, Ugan-Atik et al (11) reported this rate as 6.9%. The case with very mild dilatation of RCA was referred to our department for evaluation of end-organ damage due to hypertension, but he had no symptoms at admission and during follow-up. The other case with very mild dilatation of LAD had admitted with chest pain and further evaluation with cardiac markers, treadmill test, and CTA revealed that the chest pain was irrelevant to any cardiac disease. In our study, the rate of spontaneous closure of CPAF was 9.4% and no significant change occurred in the rest of the cases during follow-up. Most studies reported the rate of spontaneous closure of CAF as 1-2% but in contrast, Schleich et al. (17) reported this rate as 17% (1-3). Ugan-Atik et al. (11) reported the rate of spontaneous closure of CBAF in children as 3%. The considerably higher spontaneous closure rate in our study can be explained by the lack of hemodynamically significant or large fistulas. Another point is that most of the previous reports evaluated all the age groups rather than children and most studies also included all types of CAF which have a different disease course and a lower closure rate.

Although larger fistulas may be symptomatic and cause hemodynamic significance, most fistulas are usually asymptomatic. The prevalence of CPAF has increased with the advent of imaging modalities, but still there is no consensus on the

treatment of these small fistulas. Previously some authors have recommended closure of all fistulas regardless of symptoms while others recommended conservative management of small fistulas (18). The most widely-accepted treatment approach is surgical or percutaneous closure of large CPAF regardless of symptoms and closure of small fistulas in the presence of documented myocardial ischemia, arrhythmia, and otherwise unexplained ventricular dysfunction / enlargement (19). Surgical or percutaneous closure was not considered in any of the patients in our study and none of the patients had any adverse events or symptoms related to the fistula during follow-up.

Study Limitations: This study is subject to the usual limitations of a retrospective study. The number of patients with coronary dilatation, spontaneous closure, and catheter angiography is not enough to make further conclusions. Most of the data analyzed are derived from echocardiography which is an operator-dependent examination.

Coronary to pulmonary fistula is mostly an incidentally diagnosed coronary anomaly. Although the incidence of CPAF is increasing, most are small and have a benign course. They seldomly require further investigation and clinical follow-up with echocardiography is usually adequate unless they are hemodynamically significant and/or symptomatic. Patient education about preventive measures against bacterial endocarditis is of particular importance.

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Ethical Standards: The authors assert that all procedures contributing to this work comply with the ethical standards of the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional ethical committee. Informed consent was obtained from the parents of all participants

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