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Coronary artery to right pulmonary artery fistula in an adolescent

Fístula coronaria a la rama derecha de la arteria pulmonar en un adolescente: reporte de caso

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Keywords:
coronary fistula, right coronary artery, color Doppler echocardiography, multidetector-row computed tomography, right branch of the pulmonary artery.

Palabras clave:
fistula coronaria, arteria coronaria derecha, ecocardiografía Doppler color, tomografía computada multidetector, rama derecha de la arteria pulmonar.

ABSTRACT

Coronary artery fistulas (CAF) represent a rare category of congenital anomalies that are frequently underdiagnosed due to the absence of early-life symptomatology. Most cases of CAF are incidentally discovered during imaging examinations conducted for unrelated medical issues. Given the diverse presentation and potential severity of CAF progression, early screening is warranted, as irreversible cardiac remodeling may occur. Some fistulas may be large in the newborn period, but others may increase in size over time. The most substantial shunts typically occur in those fistulas where the coronary artery communicates with the right side of the heart rather than the left, which may result in symptoms of congestive heart failure, particularly in infants and occasionally in newborns. Furthermore, there have been instances of similar presentations in the elderly. Additionally, there exists a risk of thrombosis within these fistulas, which can lead to severe complications such as acute myocardial infarctions, paroxysmal fibrillation, and ventricular arrhythmias. A multimodal evaluation is crucial for achieving an accurate diagnosis at an earlier stage in life. This report presents the case of a 13-year-old female gymnast who sought medical evaluation due to palpitations.

RESUMEN

Las fístulas de las arterias coronarias (FAC) representan una categoría poco frecuente de anomalías congénitas que con frecuencia no se diagnostican debido a la ausencia de sintomatología en etapas tempranas de la vida. La mayoría de los casos de FAC se descubren de forma incidental durante un examen diagnóstico por problemas médicos no relacionados. Dada la diversidad en su presentación clínica y la posible gravedad de la progresión, se justifica la detección temprana, ya que puede producirse una remodelación cardiaca irreversible. Algunas fístulas pueden ser grandes en el periodo neonatal, pero otras pueden aumentar de tamaño con el tiempo. Los cortocircuitos significativos suelen producirse en aquellas fístulas en las que la arteria coronaria se comunica con el lado derecho del corazón en lugar del izquierdo, lo que puede provocar síntomas de insuficiencia cardiaca congestiva, en particular en lactantes y, ocasionalmente, en recién nacidos. Asimismo, ha habido casos de presentaciones similares en adultos. Además, existe un riesgo de trombosis dentro de estas fístulas, que puede provocar complicaciones graves, como infartos agudos del miocardio, fibrilación paroxística y arritmias ventriculares. La evaluación multimodal es fundamental para lograr un diagnóstico preciso en una etapa temprana de la vida. En este reporte se presenta el caso de una gimnasta de 13 años que acudió a una evaluación médica debido a palpitaciones.

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Abbreviations:
AO = Aorta
CAF = Coronary Artery Fistula
CC = Catheter Closure
CPAF = Coronary to pulmonary Artery Fistula
ECG = Electrocardiogram
LAD = left anterior descending artery
LCA = left coronary artery
LBPA = Left Branch of the Pulmonary Artery
MDCT = Multidetector-row Computed Tomography
RBPA = Right Branch of the Pulmonary Artery
RCA = Right Coronary Artery
SI = Surgical Intervention
TTE = Transthoracic Echocardiogram

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INTRODUCTION

Coronary artery anomalies encompass a group of congenital malformations characterized by alterations in the origin, course, or termination of one of the three primary epicardial arteries. Among these anomalies, coronary arteriovenous fistulas (CAF) are classified as termination anomalies. They represent abnormal direct connections between a coronary artery and a cardiac chamber, great vessel, or other vascular structures.[1] Bypassing the myocardial capillary network carries the potential to produce myocardial ischemia via the coronary steal phenomenon. The resulting left-to-right shunt may lead to volume overload, thereby increasing the risk of subsequent heart failure.[2] Coronary Arteriovenous Fistulas (CAF) are categorized as either congenital or acquired, with congenital fistulas constituting the majority of cases. Acquired CAF may develop due to intracardiac congenital heart operations, traumatic injuries to the heart, or complications arising from interventional cardiac procedures. Most presentations of CAF are isolated; however, they can also occur in conjunction with other congenital heart diseases, including atrial septal defects, ventricular septal defects, or Tetralogy of Fallot.[3] The prevalence of CAF is relatively low, occurring in approximately 0.2-0.4% of individuals with congenital heart disease. Among pediatric patients undergoing echocardiography, the prevalence is estimated at 0.06%, while in adults undergoing coronary angiography, the prevalence ranges from 0.13-0.22%.[4]

Nevertheless, advancements in imaging technology, such as multidetector-row computed tomography (MDCT), have enhanced the detection rate, leading to an overall prevalence of up to 0.9%.[5] Coronary-to-pulmonary artery fistulas represent the most common type, found incidentally on MDCT, accounting for 15-30% of the total population of CAF. Notably, 89% of these fistulas drain into the pulmonary trunk rather than into other segmental pulmonary arteries.[6]

The predominant embryologic explanation for a coronary-to-pulmonary artery fistula (CPAF) is the Hackensellner involution-persistence hypothesis. CPAF originating from the left coronary artery (LCA) represents 84% of cases, significantly outnumbering those that originate from the right coronary artery (RCA), which accounts for 38% of cases. According to the findings of Verdini et al., there are two distinct types of CPAF. The first type is characterized by a single prominent fistulous connection between either the left anterior descending artery (LAD) or the RCA and the main pulmonary trunk. The second type encompasses multiple small-caliber fistulous connections from the LAD or RCA that drain into the main pulmonary trunk.

Fistulas exhibiting a single connection are more likely to result in hemodynamic disturbances and associated symptoms compared to those with multiple connections. Although CPAF may be identified incidentally and may not be clinically significant, there exist cases with substantial hemodynamic consequences that necessitate intervention. It is important to note that the majority of CPAF have been documented solely in a limited number of case reports —there are only 22 reported pediatric cases in the existing literature— or have been discussed in relation to other congenital fistulas in larger studies.[6]

CASE PRESENTATION

A 13-year-old female gymnast presented for evaluation with complaints of palpitations, dizziness, pallor, and near-syncope. Her recorded weight was 42.7 kg (94.17 lb), height 152 cm (4 feet 11 inches), heart rate (HR) 68 beats per minute, and blood pressure (BP) 109/68 mmHg. The physical examination yielded unremarkable findings. An electrocardiogram (ECG) revealed a normal sinus rhythm without evidence of hypertrophy or alterations in repolarization. A 24-hour Holter monitor displayed normal results. The echocardiogram indicated a normal segmental relationship and chamber sizes consistent with levocardia. Both color and spectral Doppler flow assessments were normal across longitudinal and transverse views at the subcostal, parasternal, and apical levels.

The parasternal short-axis view of the aortic root showed the right coronary artery (RCA) and left coronary artery (LCA) in their appropriate anatomical positions, with the proximal segment of the RCA exhibiting a Z score of 0.2 *(Figure 1)*. However, the suprasternal longitudinal view identified an abnormal vascular structure with anterograde flow descending into the right branch of the pulmonary artery (RBPA) *(Figure 2)*. The suprasternal short-axis view illustrated an anastomosis at the proximal segment of the RBPA, characterized by continuous, pulsatile flow, a maximum recorded velocity of 2.8 m/s, and a maximum gradient of 31 mmHg *(Figure 3)*.

Based on the clinical findings and echocardiographic results, the differential diagnoses included a systemic-to-pulmonary collateral artery and/or a coronary artery fistula (CAF). However, given the normal pulmonary pressure, the coronary artery fistula was considered the primary diagnosis. A multidetector-row computed tomography (MDCT) scan was subsequently performed to establish a definitive diagnosis, revealing a coronary-to-pulmonary artery fistula originating from the sinus node artery (a branch of the proximal right coronary artery) with an anastomosis at the anterior wall of the RBPA *(Figure 4)*.

After conducting a comprehensive analysis of the case and discussing various strategies with the mother, the family concluded that the patient would continue to undergo annual reviews without pharmacotherapy.

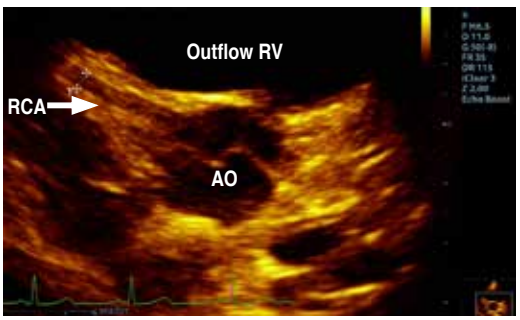

Figure 1: Parasternal short-axis 2D view from TTE, the transducer tilted superiorly and rightward in order to visualize RCA (arrow): the origin of the Right Coronary Ostium and Right Coronary Artery (RCA).

DISCUSSION

The progress in non-invasive diagnostic techniques has facilitated the early identification of incidental findings in asymptomatic pediatric patients. This advancement allows for the formulation of treatment protocols before conditions advance to irreversible cardiac remodeling. Some fistulas may be large in the newborn period, but others may increase in size over time. The most substantial shunts typically arise when a coronary artery connects to the right side of the heart instead of the left, which may result in symptoms of congestive heart failure, particularly during infancy and, on occasion, in the neonatal period. Furthermore, cases of heart failure associated with large fistulas have also been documented in the elderly population.

The occurrence of thrombosis within the fistula can lead to serious complications, including acute myocardial infarction (specifically when there is drainage to the coronary sinus) and paroxysmal atrial fibrillation (more frequently observed when the connection is to the right atrium). The most prevalent symptoms and complications observed in adults include angina, particularly in the setting of concomitant coronary artery disease. Additionally, individuals may experience myocardial infarction, heart failure, and ventricular arrhythmias.[7] Previous case reports have indicated instances of bacterial endocarditis, the formation of aneurysms, and rupture of coronary fistulas.[8]

Eighty percent of pediatric patients diagnosed with coronary artery fistulas remain asymptomatic until they reach their second decade of life. At this point, clinical symptoms may emerge, including fatigue, dyspnea, angina, and/or heart failure. A significant physical finding that typically prompts referral to a pediatric cardiologist is the detection of an asymptomatic continuous murmur over the precordium. This murmur resembles that of a patent ductus arteriosus; however, it is crucial to distinguish between the two conditions. The murmur associated with CAF is generally audible over the left lower sternal border, in contrast to the location beneath the left clavicle typically associated with murmurs originating

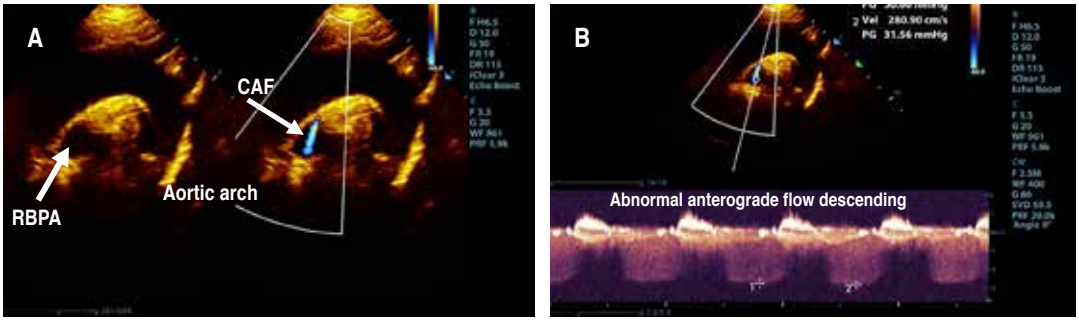

Figure 2: A) Suprasternal long-axis 2D, the transducer tilted anterior (aortic arch to the left and continuous color Doppler): view of abnormal anterograde flow descending in the RBPA (arrow). **B)** Suprasternal long-axis 2D (aortic arch to the left with continuous Doppler): view of abnormal continuous anterograde flow descending into the RBPA with Vmax: 2.8 m/s.

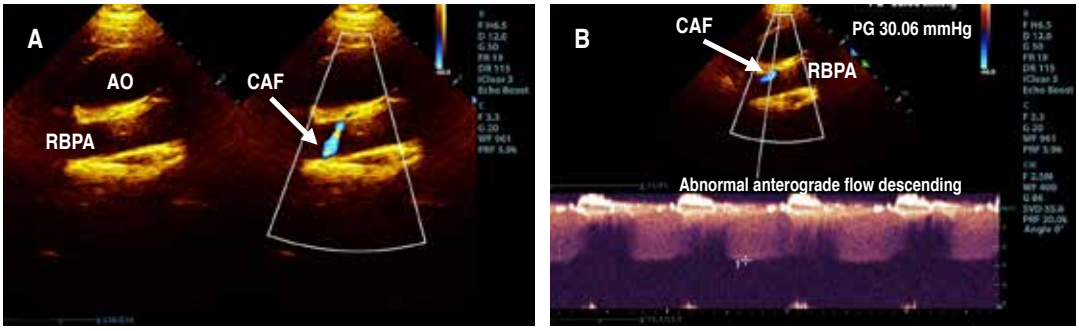

Figure 3: A) Suprasternal short-axis 2D color Doppler, the transducer tilted from anterior to posterior: view of abnormal anterograde flow descending into the proximal segment of RBPA (arrow). **B)** Suprasternal short-axis 2D with continuous Doppler: view of abnormal anterograde flow at the level of the proximal segment of RBPA with Vmax: 2.8 m/s.

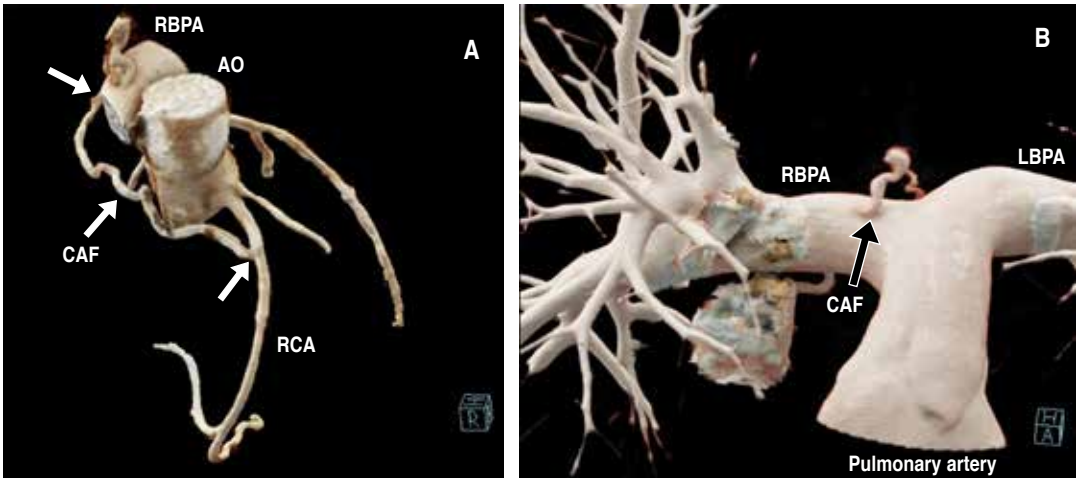

Figure 4: A) AngioTAC: view of CAF (arrow) origin at the level of the sinus node branch with distal anastomosis of the RBPA. **B)** AngioTAC: view of the CAF with distal anastomosis (arrow) in the superior wall of the proximal segment of RBPA.

from the patent arterial duct. Furthermore, the murmur typically peaks in mid-diastole rather than in systole.

Electrocardiographic findings may indicate signs of volume overload affecting both sides of the heart, as well as myocardial ischemia patterns. Chest radiographs can either appear normal or exhibit mild cardiomegaly and signs of pulmonary congestion.

Transthoracic echocardiography (TTE) may reveal CAF when the proximal coronary artery that feeds the fistula is dilated and tortuous in the presence of a large shunt. TTE is helpful in making an accurate diagnosis. In cases where the fistula and resulting shunt are small, color Doppler imaging may be diagnostic, as it effectively visualizes the chamber or vessel into which the fistula drains. Conventional pulse and continuous wave Doppler techniques can subsequently confirm the presence of high-velocity flow through the fistula.

For patients with higher body mass, multidetector computed tomography (MDCT) has proven to be more effective than echocardiography, providing precise identification of obstructions and superior anatomical delineation. Nevertheless, coronary angiography remains the gold standard for confirming the diagnosis of CAF.[9]

Congenital coronary artery anomalies present a significant risk for sudden cardiac death; however, only a select few subtypes are associated with an increased likelihood of myocardial ischemia with exertion. The most prevalent anomaly occurs when the coronary artery arises from an inappropriate sinus of Valsalva and follows an intra-arterial and intramural course. Notably, the anomalous left coronary artery originating from the right aortic sinus is recognized as carrying the highest risk, particularly among young high-endurance athletes.[10] Consequently, it is imperative to establish comprehensive screening protocols for this population.

Moreover, the clinical progression of CAF remains largely undefined, resulting in existing management guidelines being based on limited scientific evidence. According to the American College of Cardiology and the American Heart Association guidelines published in 2008, surgical intervention is classified as a class I C recommendation for large CAF, irrespective of symptomatology. For symptomatic small- to medium-sized fistulas, intervention is warranted in the presence of documented myocardial ischemia, arrhythmia, unexplained ventricular systolic or diastolic dysfunction, or endarteritis.[11] Similarly, European guidelines introduced in 2020 advocate percutaneous or surgical closure in symptomatic patients or cases involving significant shunting.[12]

The treatment options for CAF encompass both surgical intervention (SI) and catheter closure (CC). The considerable heterogeneity in size, symptomatology, and age at presentation presents a substantial challenge in determining the optimal management strategy. Recent advancements in delivery systems, microcatheters, and enhanced devices have positioned percutaneous transcatheter embolization as a safe and effective alternative in cases where anatomical conditions permit. In instances involving large aneurysmal dilations of the fistula, surgical intervention provides the opportunity for excision or reduction of the aneurysm's size.

Gowda et al. categorize CAF into two distinct types: proximal and distal. Proximal CAF originate from the central region of the proximal major epicardial artery and are generally classified as low risk for coronary events following closure because there are no normal nutritive coronary branches arising from the residual fistula segment. Conversely, distal CAF arise from the distal major epicardial coronary artery; the proximal conduit coronary artery is either tortuous or dilated and also has normal coronary artery branches supplying the myocardium.[13]

It is recommended that small-sized proximal and distal CAF undergo medical observation without intervention. All proximal CAF categorized as moderate to large, regardless of the presence of symptoms, should be closed (SI or CC) nearest to the origin of the fistula from the coronary tree as feasible, supplemented by one year of antiplatelet therapy. The decision to intervene in patients with medium to large distal CAF remains a topic of ongoing debate. Intervention at a younger age for medium-sized distal CAFs may be advisable due to favorable

remodeling mechanisms, also accompanied by one year of antiplatelet therapy.[13]

However, large distal CAF associated with significantly dilated conduit coronary arteries, irrespective of age, are considered to carry a high risk of adverse coronary events following intervention. Gowda et al. classify the treatment of larger fistulas into two distinct categories: symptomatic and asymptomatic. In cases of larger symptomatic fistulas related to heart failure, endocarditis, or hemodynamically significant runoff, the recommended approach is to undertake either SI or CC. Furthermore, it is advisable to implement a rigorous post-closure anticoagulation regimen, which should begin with intravenous heparin, followed by the administration of either warfarin or low-molecular-weight heparin for a period of six to 12 months. Additionally, antiplatelet therapy is warranted for a duration of one year or may be extended indefinitely in the presence of residual coronary dilatation.[13] The management of asymptomatic patients can be categorized into two distinct approaches. The first approach entails continuous observation accompanied by indefinite antiplatelet therapy. The second approach employs SI or CC, following the same post-closure treatment plan for large symptomatic fistulas. Regardless of the selected therapeutic approach, follow-up anatomical evaluation of the CAF at six to 12 months should elucidate remodeling sequelae that will further determine the type and duration of anticoagulant management.[13]

Mavroudis et al. advocate for elective coil occlusion in patients who meet specific criteria: absence of multiple fistulae, presence of a single narrow drainage site, absence of large branch vessels, and safe accessibility to the coronary artery supplying the fistula.[14] Therefore, early intervention in the pediatric population may be a viable option for the management of CAF, even in asymptomatic patients.[15-17]

This case presents a divergence from the existing literature, as the patient does not exhibit a murmur, a finding reported in only 12% of similar cases. Furthermore, the anastomosis was identified in the right branch of the pulmonary artery (only three cases were reported in the literature, and all were adults). Following an extensive investigation, we observed several key morphological characteristics, including a small proximal fistula and a hemodynamic circulation demonstrating a QP/QS ratio of 1.2:1. Importantly, there was no evidence to suggest that the symptoms were associated with the fistula. Consequently, the decision was made to implement annual surveillance as the appropriate management approach.

Pharmacological agents such as beta-blockers, calcium channel blockers, antiplatelet agents, or anticoagulants are often recommended for conservative management. However, many of these treatment strategies are still debated, as they are primarily founded on anecdotal evidence or a limited number of retrospective studies. A standardized protocol for the management of CPAF has yet to be established, mainly due to the infrequency of such cases and the variability in their specific anatomy and clinical presentation. Consequently, there is a pressing need for an objective tool to assess hemodynamic instability in patients with CAF.

CONCLUSIONS

Coronary artery fistulas (CAF) represent a form of heart disease that is frequently underdiagnosed, particularly during the early stages of life, as many individuals remain asymptomatic. Given the variability in presentation and the severity of CAF progression, early screening is essential to mitigate the risk of irreversible cardiovascular remodeling. A multimodal approach to evaluation facilitates the establishment of a precise diagnosis at an earlier age.

This case represents a departure from current literature, as the anastomosis is located in the right branch of the pulmonary artery, with only three such cases documented in adults to date. CPAF can be found incidentally and not be clinically significant; however, some cases can lead to substantial hemodynamic issues necessitating intervention. CPAF have been described in limited numbers of case reports or referenced within broader studies detailing other CAF. Currently, definitive treatment guidelines for CPAF remain underdeveloped. Therefore, there is a need for an objective tool to evaluate hemodynamic instability in patients presenting with CPAF.

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Ethics Statement: the authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

Consent statement: the authors declare that since this was a non-interventional, retrospective, observational study utilizing de-identified data, informed consent was not required from the patient under an IRB exemption status.

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