Scientific Research Publishing

World Journal of Cardiovascular Diseases, 2024, 14, 689-694
https://www.scirp.org/journal/wjcd
ISSN Online: 2164-5337
ISSN Print: 2164-5329

Occlusion with a Vascular Plug of a Congenital Circumflex Coronary Artery—Right Ventricle Fistula with Coronary Steal Phenomenon

Gilberto H. Acosta-Gutiérrez[1], Camila Ponce-Acosta[2], Roberto Cano-Zarate[3], Maria Jose Santa-Ana-Bayona[1], Jose A. Garcia-Montes[4], Nilda Espinola-Zavaleta[1]

[1]Nuclear Cardiology Department, National Institute of Cardiology Ignacio Chavez, Mexico City, Mexico
[2]Mexican Faculty of Medicine, La Salle University, Mexico City, Mexico
[3]Magnetic Resonance Department, National Institute of Cardiology Ignacio Chavez, Mexico City, Mexico
[4]Pediatric Interventional Cardiology Department, National Institute of Cardiology Ignacio Chavez, Mexico City, Mexico
Email: niesza2001@hotmail.com

How to cite this paper: Acosta-Gutiérrez, G.H., Ponce-Acosta, C., Cano-Zarate, R., Santa-Ana-Bayona, M.J., Garcia-Montes, J.A. and Espinola-Zavaleta, N. (2024) Occlusion with a Vascular Plug of a Congenital Circumflex Coronary Artery—Right Ventricle Fistula with Coronary Steal Phenomenon. *World Journal of Cardiovascular Diseases*, **14**, 689-694.
https://doi.org/10.4236/wjcd.2024.1411060

Received: October 17, 2024
Accepted: November 17, 2024
Published: November 20, 2024

Abstract

Background: Coronary fistulas are abnormal communications with a worldwide prevalence of 0.002% in the general population. **Aim:** The aim of this study is to present a case of a congenital coronary artery fistula presented with coronary steal and describe this phenomenon. **Methods:** A systematic search was conducted to explore the relationship between the coronary steal phenomenon and coronary artery diseases using the PubMed database. **Case Presentation:** A seven-month-old female patient, was admitted to the hospital presenting diaphoresis on feeding and failure to thrive. Physical examination denotes continuous murmur in the fourth intercostal space left parasternal border (IV/VI) crescendo-decrescendo, wide and permanent unfolding of S2 (S1-A2-P2). An echocardiogram shows enlargement of right cavities, dilatation of the left coronary artery, circumflex artery connected to the right ventricle, and pulmonary artery systolic pressure of 45 mmHg. Tomography angiography demonstrated dilatation of the circumflex artery and a tortuous trajectory towards the posterior wall of the right ventricle, suggesting a coronary fistula. **Results:** The findings were corroborated by catheterization and a vascular plug (Amplatzer AVP II) was successfully placed. During follow-up complete obliteration was observed, with adequate weight gain and no associated symptoms. **Conclusion:** This case illustrates a distinctive clinical manifestation of congenital coronary artery fistula associated with the coronary steal phenomenon, establishing a new paradigm for the early diagnosis and management of this condition in the pediatric population.

Keywords

Coronary Artery Disease, Coronary Vessel Anomalies, Embolization, Therapeutics

1. Introduction

Coronary artery fistulas (CAF) are abnormal communications between a coronary artery and any of the cardiac chambers or segments of the systemic and pulmonary circulation [1]. CAF are classified as congenital and acquired, with congenital forms being more common (90%). This condition has an estimated worldwide prevalence of 0.002% in the general population and represents 14% of congenital coronary artery anomalies [2].

The clinical relevance of this pathology is based mainly on the coronary steal phenomenon, first described in 1967 to simulate the "subclavian steal phenomenon" published by an anonymous editorial. This phenomenon describes a reversal flow in abnormal interconnected vascular networks [3]. Over time, this term has involved multiple sets of physiopathology without paraclinical confirmation of the reversal flow [4].

This study aims to present an atypical clinical case of a congenital coronary artery fistula in a lactating female, attributed to a coronary steal phenomenon.

2. Methods

A systematic search was conducted to explore the relationship between the coronary steal phenomenon and coronary artery diseases using the PubMed database. The search query employed was: ("coronary artery fistula" [Title/Abstract] OR "coronary steal phenomenon" [Title/Abstract] OR "percutaneous treatment" [Title/Abstract]). Articles were selected based on convenience and included only those published within the last 10 years.

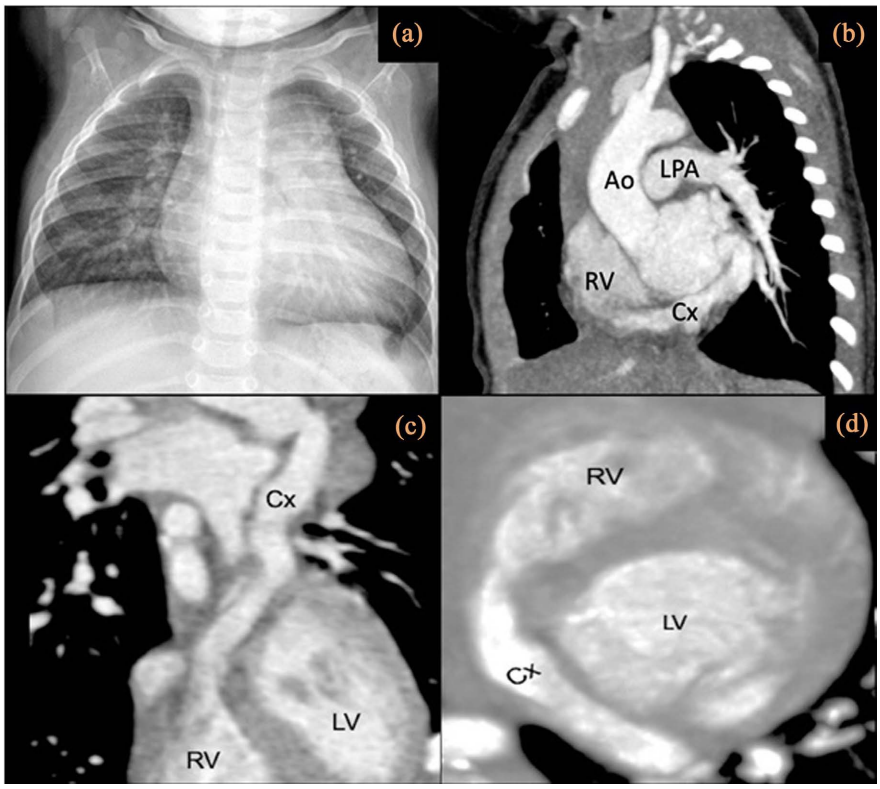
3. Case Presentation

A seven-month-old female patient with a history of systolic heart murmur since the age of two months was admitted to the institute, exhibiting diaphoresis during feeding and experiencing inadequate weight gain relative to age. In the medical history, it is significant to note that the patient is the fourth offspring, delivered prematurely at 36 weeks of gestation from a mother of advanced age. During the physical examination, vital signs were 151 bpm, 55 bpm, 36.4˚C (97.5˚F), and 94% of oxygen saturation, with a weight of 13.2 lb (1st percentile) and a height of 25.5 inches (3rd percentile). Cardiac auscultation revealed a continuous murmur in the fourth intercostal space left parasternal border (IV/VI) crescendo-decrescendo, radiated to the right hemithorax, and a wide and continued splitting of S2 (S1-A2-P2). Central and peripheral pulses were normal.

An anterior-posterior chest x-ray was conducted, which demonstrated the presence of cardiomegaly with a cardiothoracic ratio of 0.66 and pulmonary congestion (Figure 1(a)).

A transthoracic echocardiogram was performed as part of the follow-up protocol. It revealed enlargement of the right chambers and dilatation of the left main coronary artery and circumflex artery with direct flow into the right ventricle, suggesting the diagnosis of a coronary fistula. Additionally, a pulmonary artery systolic pressure of 45 mmHg was recorded, and normal systolic and diastolic biventricular function was confirmed to assess the heart failure symptoms.

Coronary computed tomography angiography (CCTA) was performed to evaluate the presence of the abnormal pathway with an infusion of 1 ml/kg of contrast medium, which showed dilatation of the circumflex artery (diameter 11.2 mm) with a tortuous path towards the posterior wall of the right ventricle, suggesting the diagnosis of circumflex coronary artery-right ventricle fistula (Figure 1(b)-(c)). These findings were corroborated by coronary catheterization (Figure 2(a)).



Abbreviations: Ao = Aorta, Cx = Circumflex coronary artery, LPA = Left Pulmonary Artery, LV = Left ventricle, RV = Right ventricle.

Figure 1. Anterior-posterior chest X-ray. Showed cardiomegaly and pulmonary congestion (Panel 1(a)). Coronary Computed Tomography Angiography presented in the sagittal section (Panel 1(b)), coronal section (Panel 1(c)), and transversal section (Panel 1(d)) reveals the presence of an abnormal connection between the circumflex coronary artery and the right ventricle.

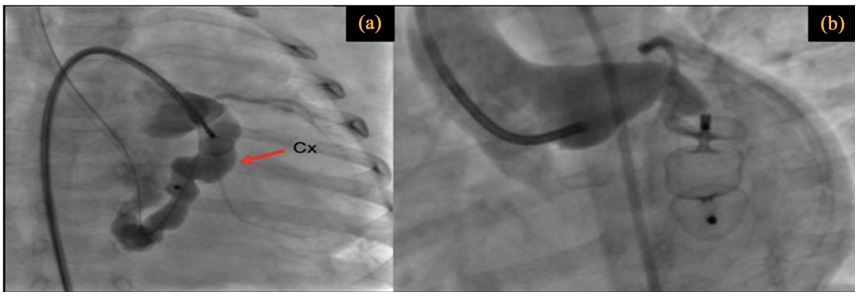


Figure 2. Coronary catheterization. The presence of a tortuous circumflex coronary artery to the right ventricle fistula was confirmed (Panel 2(a)). A percutaneous vascular plug device (Amplatzer AVP II) was successfully placed during the coronary catheterization (Panel 2(b)). Abbreviation: Cx = Circumflex coronary artery.

Due to the presence of symptoms, a percutaneous vascular plug dispositive (Amplatzer AVP II) of 12 x 9 mm was successfully placed in the coronary fistula without complications (**Figure 2(b)**), and the child was observed for 72 hours before discharge.

During follow-up, a control CCTA was performed at six months, showing a complete obliteration of the coronary fistula. Currently, the patient is asymptomatic, with adequate weight gain.

4. Discussion

Our case emphasizes the clinical manifestations of CAF and their association with the previously described "coronary steal phenomenon." It is essential to recognize that CAF are relatively rare and present with atypical clinical features. The manifestation of symptoms is predominantly linked to the resistance encountered by the fistula as it drains into the respective cardiac chambers or vessels. This resistance is influenced by three key factors: tortuosity, size, and length of the anomalous vessel [5].

The abnormal anatomical connections exhibit significant diversity; however, they can be classified into two principal categories: coronary-chamber fistulas and coronary-arteriovenous fistulas. The former represents the predominant presentation, with 55% of cases originating from the right coronary artery and 35% from the left coronary artery. The most common drainage site is the right ventricle, accounting for 41% of cases, followed by the right atrium at 26% [6]. A study conducted in Jinan, China, by Ning Li and collaborators published in 2020, with 73 patients diagnosed with CF, reported percentages of origin greater in the anterior descending artery (30%), followed by the right coronary and the circumflex artery, with 23% and 3%, respectively; despite this change in the frequency, the least common artery presentation is the one observed in the case [7].

The clinical presentation of coronary artery fistulas in infants differs significantly from that in patients during the second decade of life. Most infants are asymptomatic; however, in our case, symptoms of heart failure were evident, including failure to thrive, fatigue during feeding, diaphoresis, tachycardia, and tachypnea. A characteristic finding during auscultation was the presence of a

continuous murmur (systolic-diastolic) accompanied by an increased second heart sound or persistent splitting of S2. This murmur typically exhibits a crescendo-decrescendo pattern [8].

Particularly, in this patient, diaphoresis on feeding and failure to thrive could be explained by the coronary steal phenomenon, a decreased flow of the coronary artery due to abnormal communication [3]. The coronary blood flow depends on two main factors: aortic pressure and coronary fistula resistance. In stress situations, the oxygen flow increases in the myocardium due to the vasomotor process, reducing the coronary resistance. This reduction of arterial resistance in the presence of a coronary fistula leads to a decrease in the myocardial perfusion pressure, especially distal to the origin of the fistulous connection [9].

For the imaging evaluation, the transthoracic echocardiogram has presented important limitations; it has been reported that it presents low accuracy for the diagnosis of CAF, with few reports in infants, being observed 19.1% in a study with 72 adult patients [10]. Despite that, the assessment of ventricular and atrial function and the search for pulmonary hypertension are essential to establish a specific treatment [11]. Consequently, the imaging modality of choice for evaluating CAF is CCTA. However, despite its high precision, the presence of distal dilatations, which are common in children, may not be observed due to the loss of vision in these distal segments of the coronary arteries, which has repercussions for surgical management [10].

Currently, the therapeutic management of coronary fistulas remains a subject of debate. In cases of asymptomatic fistulas, expectant management is generally preferred, while symptomatic presentations typically warrant percutaneous closure as the first-line treatment, as was the case with our patient. Surgical correction options include epicardial ligation, transection, and closure of the drainage opening [12]. In recent years, a novel embolization technique using vascular plugs has emerged as a less invasive and cost-effective alternative, associated with reduced recovery times and lower mortality rates compared to the traditional surgical closure method. In 2015, Ilkay and colleagues conducted a follow-up study involving 20 patients who underwent vascular plug occlusion for CAF, with evaluations at 6 months showing no vascular complications [13].

Overall, these evolving therapeutic strategies highlight the need for individualized treatment approaches based on clinical presentation and patient-specific factors.

5. Conclusion

This case highlights a unique instance in an infant who typically presents asymptomatically, exhibiting symptoms related to a coronary steal phenomenon. The diagnosis and management were confirmed through percutaneous coronary catheterization, underscoring the need for further investigation into the diagnostic and therapeutic approaches for this condition in the pediatric population.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the

ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975.

Permission

Permission for presentation has been obtained from the patient's parents.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Kastellanos, S., Aznaouridis, K., Vlachopoulos, C., Tsiamis, E., Oikonomou, E. and Tousoulis, D. (2018) Overview of Coronary Artery Variants, Aberrations and Anomalies. *World Journal of Cardiology*, **10**, 127-140. https://doi.org/10.4330/wjc.v10.i10.127

[2] Buccheri, D. (2021) Fístulas da Artéria Coronária. *Arquivos Brasileiros de Cardiologia*, **117**, 89-90. https://doi.org/10.36660/abc.20210501

[3] Johnson, N.P., Kirkeeide, R.L. and Gould, K.L. (2021) Coronary Steal: Mechanisms of a Misnomer. *Journal of the American Heart Association*, **10**, e021000. https://doi.org/10.1161/jaha.121.021000

[4] Vaidya, Y.P. and Green, G.R. (2019) Coronary Artery Fistula. *Journal of Cardiac Surgery*, **34**, 1608-1616. https://doi.org/10.1111/jocs.14267

[5] Huang, C. and Deng, L. (2024) Left Coronary Artery–to–Right Coronary Sinus Fistula. *Radiology*, **310**, e233027. https://doi.org/10.1148/radiol.233027

[6] Ramos, D.d.O.S., Sarmet dos Santos, A.R., Vaz, A. and Ishikawa, W.Y. (2023) Giant Congenital Coronary-Cameral Fistula. *Radiology*: *Cardiothoracic Imaging*, **5**, e220245. https://doi.org/10.1148/ryct.220245

[7] Li, N., Zhao, P., Wu, D. and Liang, C. (2019) Coronary Artery Fistulas Detected with Coronary CT Angiography: A Pictorial Review of 73 Cases. *The British Journal of Radiology*, **93**, Article 20190523. https://doi.org/10.1259/bjr.20190523

[8] Velázquez-Castañeda, R., De La Puente-Tawil, I., Molina-Castillo, S., Proaño-Bernal, L., Rodríguez-Salazar, M.A., Masso-Bueso, S., *et al*. (2023) Coronary-Cameral Fistula in an Infant Patient: An Incidental Diagnosis. *Clinical Case Reports*, **11**, e8172. https://doi.org/10.1002/ccr3.8172

[9] Achim, A., Johnson, N.P., Liblik, K., Burckhardt, A., Krivoshei, L. and Leibundgut, G. (2023) Coronary Steal: How Many Thieves Are Out There? *European Heart Journal*, **44**, 2805-2814. https://doi.org/10.1093/eurheartj/ehad327

[10] Goo, H.W. (2021) Imaging Findings of Coronary Artery Fistula in Children: A Pictorial Review. *Korean Journal of Radiology*, **22**, 2062-2072. https://doi.org/10.3348/kjr.2021.0336

[11] Erdogan, E. and Kilic, Y. (2022) An Atypical Cause of Chest Pain. *Cardiology in the Young*, **32**, 1839-1841. https://doi.org/10.1017/s1047951122000476

[12] Bhasin, D., Arora, G., Giridhara, P., Gupta, A., Isser, H.S. and Bansal, S. (2021) Coronary-Cameral Fistula to Pulmonary Artery: An Innocent Bystander? *Journal of Invasive Cardiology*, **33**, E70. https://doi.org/10.25270/jic/20.00108

[13] Ilkay, E., Celebi, O.O., Kacmaz, F. and Ozeke, O. (2015) Percutaneous Closure of Coronary Artery Fistula: Long-Term Follow-Up Results. *Advances in Interventional Cardiology*, **4**, 318-322. https://doi.org/10.5114/pwki.2015.55603