Revista MedEs
Revista Científica Estudiantil de Matanzas
MedEst. 2025 Vol.5: e368 ISSN: 2789-7567 RNPS: 2524

CASE PRESENTATION

Surgical approach and rehabilitation in ARCAPA syndrome: report of a pediatric case

Abordaje quirúrgico y rehabilitación en síndrome ARCAPA: reporte de un caso pediátrico

Miguel Enrique Barroso Fontanals [1]*, https://orcid.org/0000-0003-3291-7457

Karla María Ascanio Cruz [2], https://orcid.org/0000-0002-2169-8618

Carlos Roberto Urgelles Oliva [1], https://orcid.org/0009-0000-7576-1427

Malena Laura Vázquez Núñez [3], https://orcid.org/0009-0000-4357-6171

[1] University of Medical Sciences of Santiago de Cuba. Faculty No. 2 of Medicine. Santiago de Cuba, Cuba.

[2] University of Medical Sciences of Santiago de Cuba. Faculty No. 1 of Medicine. Santiago de Cuba, Cuba.

[3] Matanzas University of Medical Sciences. Dr. Juan Guiteras Gener Faculty of Medical Sciences of Matanzas. Matanzas, Cuba.

*** Corresponding author:** mbarrosof9@gmail.com

Received: 24/06/2025

Accepted: 06/09/2025

How to cite this article: Barroso-Fontanals ME, Ascanio-Cruz KM, Urgelles-Oliva CR, Vázquez-Núñez ML. Surgical approach and rehabilitation in ARCAPA syndrome: report of a pediatric case. MedEst. [Internet]. 2025 [cited access date]; 5:e368. Available in: https://revmedest.sld.cu/index.php/medest/article/view/368

ABSTRACT

MedEst. 2025 Vol.5; e368
ISSN: 2789-7567 RNPS: 2524

Introduction: anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA syndrome) is an extremely rare congenital malformation, with an estimated prevalence of 0.002% among all congenital heart defects. This condition is characterized by an anomalous origin of the right coronary artery from the pulmonary trunk, resulting in reverse coronary flow, risk of myocardial ischemia, and life-threatening complications, especially in the pediatric population.

Objective: to describe the case of a pediatric patient diagnosed with ARCAPA syndrome.

Case presentation: a 15-year-old male adolescent with no significant medical history presented with fatigue, anginal-like chest pain, and dyspnea on exertion. Physical examination revealed muffled heart sounds and asynchronous peripheral pulses. The electrocardiogram showed changes suggestive of ischemia, and cardiac magnetic resonance imaging confirmed the anomalous origin of the right coronary artery from the pulmonary artery with retrograde flow. Surgical correction was performed with an end-to-side anastomosis to the ascending aorta, followed by a cardiac rehabilitation program. The outcome was favorable, with complete restoration of cardiovascular function.

Conclusions: this condition is a potential cause of sudden death in young patients and should be included in the differential diagnosis of chest pain and exertional dyspnea. Accurate detection requires Doppler echocardiography and cardiac magnetic resonance imaging. Treatment with early surgical anastomosis restores coronary flow, complemented by cardiac rehabilitation. Multidisciplinary management involving cardiologists, surgeons, and rehabilitation specialists is essential for the therapeutic success of this complex congenital anomaly.

Keywords: Cardiovascular Anomalies; Congenital Heart Disease; Cardiac Surgical Procedures; Cardiac Rehabilitation; Pulmonary Artery

RESUMEN

Introducción: el origen anómalo de la arteria coronaria derecha desde la arteria pulmonar (síndrome de ARCAPA), es una malformación congénita extremadamente rara, con una prevalencia estimada del 0,002 % entre todas las cardiopatías congénitas. Esta condición se caracteriza por un origen anómalo de la arteria coronaria derecha en el tronco pulmonar, lo que provoca

MedEst. 2025 Vol.5; e368
ISSN: 2789-7567 RNPS: 2524

un flujo coronario invertido, riesgo de isquemia miocárdica y complicaciones potencialmente mortales, especialmente en población pediátrica.

Objetivo: describir el caso de un paciente pediátrico diagnosticado con síndrome de ARCAPA.

Presentación del caso: adolescente masculino de 15 años sin antecedentes de interés, que acudió por astenia, dolor torácico de características anginosas y disnea de esfuerzo. El examen físico reveló ruidos cardíacos apagados y pulsos periféricos asincrónicos. El electrocardiograma mostró alteraciones sugerentes de isquemia, y la resonancia magnética cardíaca confirmó el origen anómalo de la arteria coronaria derecha desde la arteria pulmonar con flujo retrógrado. Se realizó corrección quirúrgica mediante anastomosis término-lateral a la aorta ascendente, seguida de un programa de rehabilitación cardiaca. La evolución fue favorable, con restablecimiento completo de la función cardiovascular.

Conclusiones: esta entidad es una causa potencial de muerte súbita en jóvenes que debe incluirse en el diagnóstico diferencial de dolor torácico y disnea de esfuerzo. Su detección precisa requiere ecocardiograma Doppler y resonancia magnética cardíaca. El tratamiento con anastomosis quirúrgica temprana restaura el flujo coronario, complementado con rehabilitación cardiaca. El manejo multidisciplinario entre cardiólogos, cirujanos y especialistas en rehabilitación es fundamental para el éxito terapéutico de esta anomalía congénita compleja.

Palabras Clave: Anomalías Cardiovasculares; Cardiopatías Congénitas; Procedimientos Quirúrgicos Cardíacos; Rehabilitación Cardiaca; Arteria Pulmonar

INTRODUCTION

Anomalous coronary artery originating from the pulmonary artery (ARCAPA) syndrome is an extremely rare congenital malformation characterized by the anomalous origin of a coronary artery from the pulmonary trunk instead of the aorta. This condition results in inadequate myocardial blood supply, which can lead to ischemia, serious complications, and even sudden death, especially in the pediatric population, if not promptly diagnosed and treated. (1)

Initially described in the mid-20th century, its detection has been revolutionized by advances in cardiac imaging techniques, allowing for accurate characterization and early management. At the same time, the

refinement of surgical techniques has significantly improved patient prognosis. (2,3)

With an estimated prevalence of approximately 0,002 % of all congenital heart defects, it is usually diagnosed in childhood or adolescence with the onset of symptoms such as dyspnea, chest pain, or fatigue during exercise, and is frequently associated with coronary aneurysms and fistulous tracts. (4,5)

Due to its rarity and clinical relevance, this article aims to describe the case of a pediatric patient diagnosed with ARCAPA Syndrome.

CASE PRESENTATION

A 15-year-old male patient from an urban setting with no significant personal or family history presented with asthenia, anginal-like chest pain, and exertional dyspnea of several weeks' duration.

Physical Examination:

The patient was in good general condition, conscious, oriented, and normohydrated. Vital signs showed a heart rate of 88 bpm, blood pressure of 110/70 mmHg, respiratory rate of 16 bpm, and oxygen saturation of 98%. Cardiovascular examination revealed rhythmic but muffled heart sounds, no audible murmurs, present but asynchronous peripheral pulses, and no jugular vein engorgement. Respiratory examination revealed preserved vesicular murmurs without any accompanying sounds. The remainder of the physical examination revealed no relevant findings.

Complementary Tests:

Electrocardiogram: Slightly widened QRS complex with sporadic ventricular ectopics, suggestive of conduction disturbances or underlying ischemia.

Cardiac MRI: Anomalous origin of the right coronary artery from the main pulmonary artery, with aneurysmal dilation of the vessel and retrograde flow evident on contrast sequences.

Doppler echocardiogram confirmed an anomalous course of the right coronary artery and suggested reversed coronary flow.

After a thorough medical evaluation, the diagnosis of ARCAPA syndrome was confirmed. The patient was transferred to the Cardiocenter of Santiago de Cuba for surgical correction. The procedure consisted of a midline thoracotomy

MedEst. 2025 Vol.5; e368
ISSN: 2789-7567 RNPS: 2524

with dissection of the right coronary artery at its pulmonary origin and subsequent end-to-side anastomosis to the ascending aorta, thus restoring adequate coronary flow.

The postoperative period was uneventful, with close monitoring of cardiac function, which remained stable. A supervised cardiovascular rehabilitation program was implemented, allowing for progressive recovery. The patient showed favorable progress, with significant symptomatic improvement and complete restoration of functional capacity without residual limitations.

Short- and medium-term follow-up demonstrated graft patency and absence of complications, confirming the success of the therapeutic approach.

DISCUSSION

ARCAPA syndrome is rare, accounting for only 0,002 % of all congenital heart defects. Landi et al. [6] report that most patients are asymptomatic and the anomaly is detected in adulthood. This is inconsistent with the case presented here, as the patient was in a pediatric age group and symptomatic.

Occasionally, ARCAPA can cause myocardial ischemia and/or sudden cardiac arrest. Some studies [7,8] report that surgical correction is recommended in most patients, with the goal of eliminating the left-to-right shunt and establishing dual coronary circulation to prevent the potential risk of myocardial ischemia due to coronary steal. The patient underwent cardiovascular revascularization surgery to correct this anomaly. During the procedure, the right coronary artery was reconnected to the aorta to restore adequate blood flow to the heart.

To suspect ARCAPA syndrome in a patient, a thorough clinical evaluation is crucial, including a thorough physical examination, complementary tests, and cardiac imaging tests to confirm the diagnosis. Castro et al. [9] indicate that several imaging techniques are available for the diagnosis of coronary anomalies, such as echocardiography, coronary angiography, magnetic resonance imaging, and coronary arteriography, but coronary computed tomography is the gold standard.

If there is any suspicion, evaluation by a specialized cardiologist is vital for the appropriate approach and management of this congenital heart condition. [10,11] In this patient, the use of magnetic resonance imaging is justified, despite its secondary role in these pathologies, due to the patient's age, and high doses of radiation are avoided.

Cardiac rehabilitation is a fundamental pillar in the postoperative management of patients with ARCAPA syndrome, as demonstrated in the case presented. Although it has traditionally focused on adult ischemic heart disease, current evidence supports its usefulness in the pediatric population undergoing surgical correction of congenital coronary anomalies. Khajali et al. [13] highlight how these programs optimize functional recovery and prevent long-term complications through structured monitoring, supervised exercise, and therapeutic education. The timely implementation of cardiac rehabilitation in our patient allowed not only the restoration of his functional capacity but also a gradual return to his activities without residual limitations, corroborating its value in the therapeutic spectrum of this condition.

CONCLUSIONS

ARCAPA syndrome is a rare but potentially fatal entity in young people, which should be considered when chest pain and exertional dyspnea occur. Accurate diagnosis requires Doppler echocardiography and cardiac magnetic resonance imaging. Early surgical correction by aortic anastomosis has proven effective in restoring physiological coronary flow. Postoperative cardiac rehabilitation and a multidisciplinary approach involving cardiologists, surgeons, and rehabilitation specialists are essential for the successful management of this complex congenital anomaly.

BIBLIOGRAPHIC REFERENCES

1. Franco A, Cadenasso B, Antúnez S, Anzíbar R, Touyá G. Origen anómalo de la arteria coronaria derecha desde el tronco de la arteria pulmonar (ARCAPA). Arch. Pediatr. Urug. [Internet]. 2018 [cited 10/06/2025]; 89(2): 108-112. Available in: http://www.scielo.edu.uy/scielo.php?script=sci_arttext&pid=S1688-12492018000200108&lng=es

2. Kolesár A, Toporcer T, Čobejová J, Lukačin Š. Anomalous Arising of Right Coronary Artery from the Pulmonary Artery. Journal of Cardiovascular Development and Disease [Internet]. 2024 [cited 10/06/2025]; 11(2): 50. Available in: https://doi.org/10.3390/jcdd11020050

3. Arqué Gibernau JM, Arias Recalde A, Bravo Marqués R. Síndrome de ARCAPA en la edad adulta. Rev Esp Cardiol [Internet]. 2017 [cited 10/06/2025]; 70(8): 665. Available in: https://www.revespcardiol.org/es-sindrome-arcapa-edad-adulta-articulo-S0300893216304390

4. Suchodolski A, Galeczka M, Szulik, Fiszer R, Glowacki J. Anomalous origin of the right coronary artery originating from the pulmonary trunk: pre-and postoperative computed tomography images and virtual reality reconstructions. Coronary Artery Disease [Internet]. 2024 [cited 10/06/2025]; 35(2): 164 - 165. Available in: https://doi.org/10.1097/MCA.0000000000001315

5. Karila-Cohen J, Laux D, Houyel L, Bonnet D. Aorta without coronary arteries: anatomic variants of a rare malformation. Pediatric Cardiology [Internet]. 2024 [cited 10/06/2025]; 45(1): 200 - 207. Available in: https://doi.org/10.1007/s00246-023-03330-7

6. Landi I, Alkhabaz A, Abou Shaar B, Galzerano D, Albert-Brotons D, Tahir M, et al. Non-atherosclerotic coronary artery disease: an overview of a heterogeneous disease. Coronary Artery Disease [Internet]. 2024 [cited 10/06/2025]; 10: 1097. Available in: https://doi.org/10.1097/MCA.0000000000001317

7. Song Y, Choi ES, Kim DH, Kwon BS, Park CS, Yun TJ. Surgical Managements of Coronary Artery Fistulas in Children.Journal of Chest Surgery [Internet]. 2024 [cited 10/06/2025]; 57(1): 79. Available in: https://doi.org/10.5090/jcs.23.101

8. Giamberti A, Chessa M, Evangelista M, Caldaroni F. Anomalous origin of the Coronary Arteries from the Pulmonary Artery: ALCAPA and ARCAPA. Congenital Anomalies of Coronary Arteries [Internet]. 2023 [cited 10/06/2025]; 75 - 83. Available in: https://doi.org/10.1007/978-3-031-36966-7_7

9. Castro CR, Fachado F. Síndroma ARCAPA: o papel do médico de familia na gestão de uma doença rara. Revista Portuguesa de Meicina Geral e Familiar [Internet]. 2023 [cited 10/06/2025]; 39(1): 67 - 72. Available in: https://doi.org/10.32385/rpmgf.v39i1.13470

10. González Hoyos D, López Arroyave M, Sebastián Quiñones J, Abad Díaz P, Carvajal Vélez M. Anomalías de las arterias coronarias: una revisión de la literatura y propuesta de una nueva clasificación. Revista Colombiana de Cardiología. [Internet] 2023 [cited 10/06/2025]; 30(6): 390 – 402. Available in: http://scielo.org.co/scielo.php?script=sci_arttext&pid=S0120-56332023000600390&Ing=en

11. Fernández Cordón C, Prieto Arévalo R, Bermejo J, Fernández Avilés F. Actualización en cardiopatías congénitas del adulto. Medicine-Programa de Formación Médica Continuada creditado. [Internet] 2021 [cited 10/06/2025]; 13(41): 2379 – 2389. Available in: https://sciencedirect.com/science/article/pii/S0304541221002353
12. Faz Pujalte M, Richart Martínez M, Perpiñá Galván J. Análisis de la efectiviad de la rehabilitación cardiaca en España: una revisión istematica exploratoria. Anales del Sistema Sanitario de Navarra [Internet] 2022 [cited 10/06/2025]; 45(1). Available in: http://scielo.isciii.es/scielo.php?script=sci_arttext&pid=S1137-66272022000100011&Ing=en

13. Khajali Z, Zangenehfar ME, Maadani M, Parsaee M. A Young Woman with Atypical Chest Pain: Introducing a rare Case of ARCAPA. Multidisciplinary Cardiovascular Annals [Internet]. 2024 [cited 10/06/2025]; 11(2). Available in: https://doi.org/10.51812/mca.106461

STATEMENT OF AUTHORSHIP

MEBF: project administration, conceptualization, data curation, research, methodology, resources, supervision, validation, visualization, writing - original draft, writing - review and editing.

KMAC: conceptualization, data curation, research, methodology, validation, visualization, writing - original draft.

CRUO: conceptualization, research, validation, writing - review and editing.

MLVN: conceptualization, research, validation, writing - review and editing.

CONFLICT OF INTERESTS

The authors have no conflicts of interest to declare.

SOURCES OF FUNDING

The authors did not receive funding for the development of this article.