Early Surgical Management of Neonatal Coronary Artery Fistula with Severe Hemodynamic Impact

Catarina Cezanne Isabel Graça Tomas Sim Sim Carla Saraiva Ana Rita Araújo João Rato Marta Marques Mónica Rebelo

PII: S0870-2551(25)00392-0

DOI: https://doi.org/doi:10.1016/j.repc.2025.09.009

Reference: REPC 2518

To appear in: *Revista Portuguesa de Cardiologia*

Received Date: 5 April 2025

Accepted Date: 20 September 2025

Please cite this article as: Cezanne C, Graça I, Sim TS, Saraiva C, Araújo AR, Rato J, Marques M, Rebelo M, Early Surgical Management of Neonatal Coronary Artery Fistula with Severe Hemodynamic Impact, *Revista Portuguesa de Cardiologia* (2025), doi: https://doi.org/10.1016/j.repc.2025.09.009

Title: Early Surgical Management of Neonatal Coronary Artery Fistula with Severe Hemodynamic Impact

Título: Tratamento cirúrgico precoce de fístula arterial coronária neonatal com significado hemodinâmico grave

First coauthors:

- Catarina Cezanne*, Serviço de Pediatria, ULS Almada-Seixal, Almada, Portugal

- Isabel Graça, Serviço de Cardiologia Pediátrica, ULS Lisboa Ocidental, Lisboa, Portugal

Other authors:

- Tomas Sim Sim, Serviço de Cardiologia Pediátrica, ULS Lisboa Ocidental, Lisboa, Portugal

- Carla Saraiva, Serviço de Radiologia, ULS Lisboa Ocidental, Lisboa, Portugal

- Ana Rita Araújo, Serviço de Cardiologia Pediátrica, ULS Lisboa Norte, Lisboa, Portugal

- João Rato, Serviço de Cardiologia Pediátrica, ULS Lisboa Ocidental, Lisboa, Portugal

- Marta Marques, Serviço de Cirurgia Cardiotorácica, ULS Lisboa Ocidental, Lisboa, Portugal

- Mónica Rebelo, Serviço de Cardiologia Pediátrica, ULS Lisboa Norte, Lisboa, Portugal

***Corresponding author**:

E-mail address: catarinacezanne@gmail.com (C. Cezanne)

A term newborn (38weeks, 2965g) was diagnosed prenatally with a large coronary artery fistula (CAF) from the left anterior descending artery to the apex of the right ventricle, causing hemodynamic compromise, including right ventricular dilation and retrograde aortic flow (Figure1). The delivery was planned to take place at a tertiary center with cardiac assistance. After an uneventful vaginal delivery, the baby was transferred for surgical intervention.

Echocardiography confirmed a large CAF with bidirectional flow and systemic diastolic steal. Angio-CT showed a large, tortuous vessel (8x7mm at origin), distal diameter of 6 mm, with significant dilation of the right ventricle and atrium(Figure2). Surgical ligation of the fistula was performed on day 3 (Figure2). Persistent fistula on follow-up echo and Angio-CT required re-ligation above the previous site on day 11, which resolved the condition. Both surgeries were uneventful, performed without extracorporeal circulation. The baby was discharged on day 17 with anticongestive therapy and is currently doing well, weighing 4600g at 2 months.

Coronary artery fistula is a rare congenital anomaly (1 in 50 000 live births), involving abnormal connections between coronary arteries and cardiac chambers (1-5). The prenatal diagnosis is complex. Although it is usually asymptomatic, large or symptomatic

CAFs may cause heart failure or ischemia and often require intervention. While transcatheter closure is preferred for its safety(3), surgical repair is necessary for complex cases (2,4,5). This case highlights the importance of prenatal diagnosis, allowing for delivery planning at a referral center and early intervention, which improves morbidity (1,5). Follow-up is essential to check ventricular and vascular remodeling and to prevent complications(1,4,5).

Funding: none.

All authors have read and approved the manuscript.

Declarations of interest: none.

Ethics in publishing

1. Does your research involve experimentation on animals?:

No

2. Does your study include human subjects?:

No

3. Does your study include a clinical trial?:

No

4. Are all data shown in the figures and tables also shown in the text of the Results section and discussed in the Conclusions?:

Yes

References:

1. Scalera S, Clemente A, Pizzuto A, et al. 3D Printed Model-Guided Neonatal Transcatheter Closure of Left Main Coronary Artery-to-Right Ventricle Fistula. *J Am Coll Cardiol Case Rep*. 2023;16:101869.

2. Ismail AQT, Gandhi A, Desai T, et al. A Neonatal Case of Congenital Coronary Artery Fistula. *BMJ Case Rep*. 2012;2012:bcr.09.2011.4773.

3. Aggarwal V, Mulukutla V, Qureshi AM, et al. Congenital Coronary Artery Fistula: Presentation in the Neonatal Period and Transcatheter Closure. *Congenit Heart Dis*. 2018;1–6.

4. Buccheri, D., Chirco, P. R., Geraci, S., et al. (2018). Coronary Artery Fistulae: Anatomy, diagnosis and Management Strategies. *Heart Lung and Circulation*, *27*(8), 940–951. https://doi.org/10.1016/j.hlc.2017.07.014

5. Torres, S., Vasconcelos, M., Silva, M. T., et al. (2022). Coronary artery fistulas: A 12-year single-center experience. *Revista Portuguesa De Cardiologia*, *41*(10), 843–850. https://doi.org/10.1016/j.repc.2021.06.024

Figures

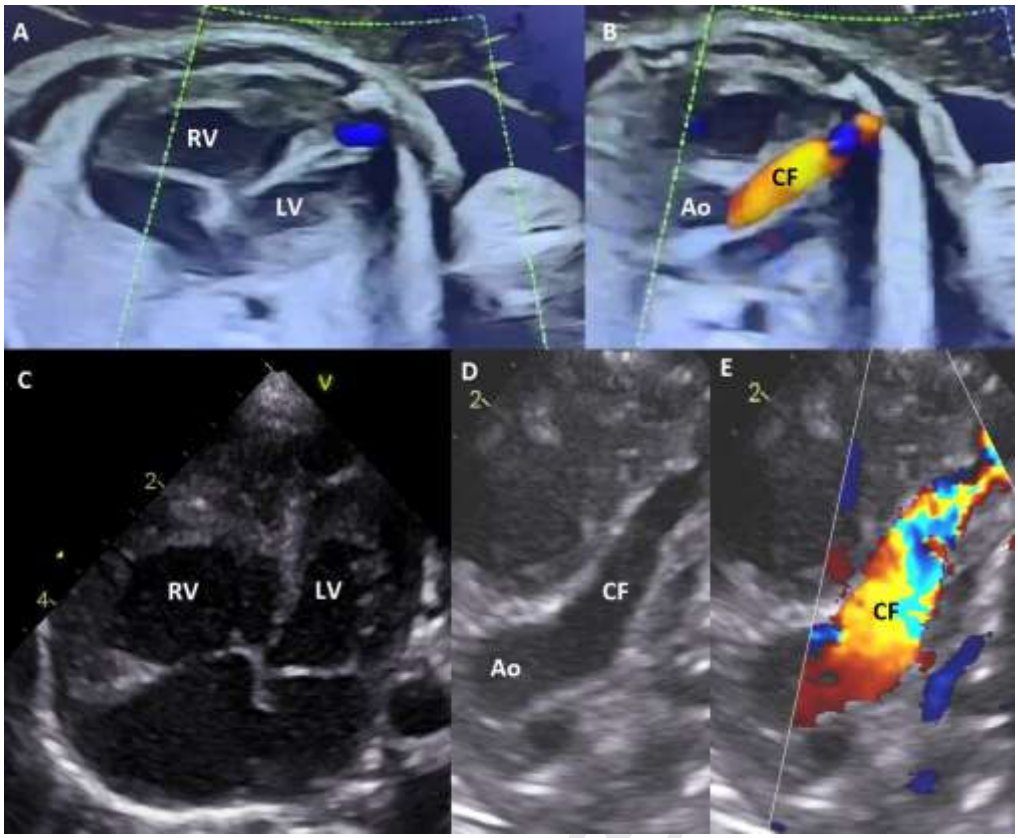

Figure 1: Prenatal and postnatal echocardiograms showing right ventricular dilatation and large coronary fistula. A – Prenatal 4-chamber view with right ventricular dilatation, compared to the left ventricle. B – Prenatal view showing a large coronary fistula with antegrade flow from the aorta to the apex of the right ventricle. C – Postnatal 4-chamber view with right ventricular dilatation. D and E – Postnatal view showing a large coronary fistula with antegrade flow from the aorta. Legend: Ao: aorta; CF: coronary fistula; LV: left ventricle; RV: right ventricle.

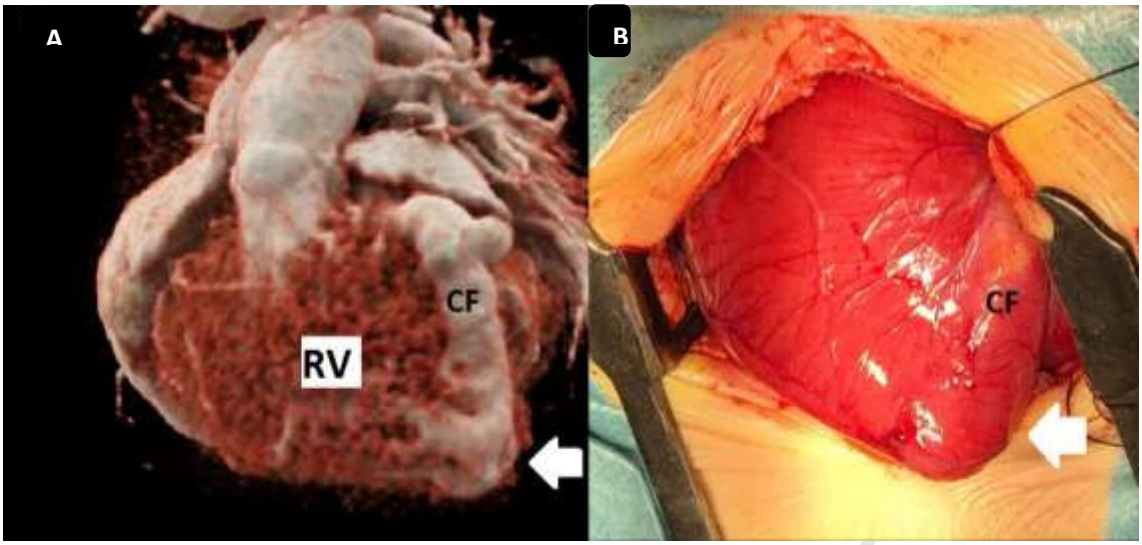

Figure 2: A: computed tomography angiography (CT-angio) three dimensional images showing the whole fistula to its distal end as it enters the right ventricular cavity (white arrow). B - The same view from the surgeon's perspective. Multimodal evaluation allows for excellent preoperative assessment and adequately represents the anatomy the surgeon will find.