



# Multimodality imaging in complex aortic arch anomaly

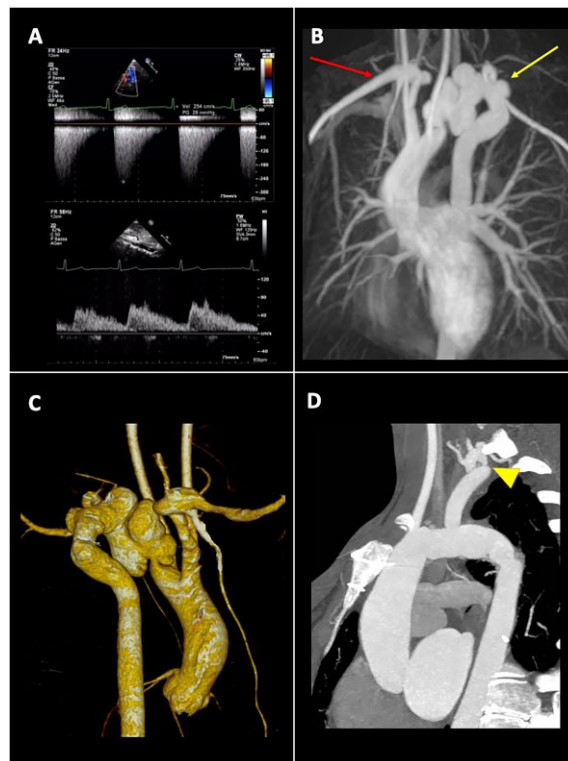
**Gaia Spaziani** <sup>1\*</sup>, **Elena Bennati**<sup>1</sup>, **Silvia Favilli**<sup>1</sup>, **Paolo Guccione**<sup>2</sup>, and **Aurelio Secinaro** <sup>3</sup>

<sup>1</sup>Pediatric Cardiology Unit, Department of Pediatrics, Meyer Children's Hospital, Viale Gaetano Pieraccini 24, 50139 Florence, Italy; <sup>2</sup>Pediatric Cardiology and Cardiac Surgery Department, Bambino Gesù Children's Hospital IRCCS, Piazza Sant'Onofrio 4, 00165 Rome, Italy; and <sup>3</sup>Department of Imaging, Bambino Gesù Children's Hospital IRCCS, Piazza Sant'Onofrio 4, 00165 Rome, Italy

Received 16 November 2021; first decision 23 November 2021; accepted 17 January 2022; online publish-ahead-of-print 3 February 2022

A 5-year-old female was referred to our centre for evaluation of a cardiac murmur. Transthoracic echocardiography showed a cervical aortic arch (AA) with mild flow acceleration at aortic isthmus. Owing

to a suspicion of aortic coarctation (CoA), a computed tomography (CT) was performed, revealing a dysmorphic and tortuous S-shaped left AA without signs of CoA. The patient underwent an annual



\* Corresponding author. Tel: +39 0555662498, Email: [gaiaspaziani@gmail.com](mailto:gaiaspaziani@gmail.com)

Handling Editor: Michel Pompeu Sá

© The Author(s) 2022. Published by Oxford University Press on behalf of the European Society of Cardiology.

This is an Open Access article distributed under the terms of the Creative Commons Attribution-NonCommercial License (<https://creativecommons.org/licenses/by-nc/4.0/>), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact [journals.permissions@oup.com](mailto:journals.permissions@oup.com)



**Video 1** Baseline angio-CMR of aorta showed multiple kinking and aortic arch dilatation.

follow-up. At the age of 14 years, echocardiographic findings consistent with the diagnosis of CoA were detected (diastolic run-off at aortic isthmus and demodulated pattern flow in the abdominal aorta; *Panel A*). Cardiovascular magnetic resonance (CMR) was performed, showing multiple kinking, AA dilation, and proximal right subclavian artery ectasia (*Panel B*, yellow and red arrows, respectively; *Video 1*). Genetic analysis showed pathogenetic mutation in transforming growth factor- $\beta$ 2 gene, associated to Loeys-Dietz syndrome. Two years later, CMR showed a worsening of AA findings with maximum dilation of 40 mm with obvious flow acceleration just proximal to the aneurysmal site. Following a new CT (*Panel C*; *Video 2*), the patient was scheduled for an intervention of AA reconstruction with conduit; moreover, an additional conduit was implanted at the left sub-



**Video 2** Preoperative angio-CT showed a worsening of kinking and aortic arch dilatation.

clavian artery origin. Six months later, a CT revealed harmonious repaired AA morphology with only discrete stenosis at subclavian artery-conduit anastomosis (*Panel D*, yellow arrowhead), the latter was successfully treated percutaneously.

Genetic aortopathies are complex conditions requiring a multidisciplinary approach.

Multimodality imaging is of paramount importance in the management of aortopathies, from diagnosis to follow-up and it may guide timing and type of treatment.

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** None declared.

**Funding:** None declared.