JACC: CASE REPORTS VOL. 29, 2024

CONGENITAL HEART DISEASE

CASE REPORT: MULTIDISCIPLINARY TEAM DISCUSSIONS

Percutaneous Coronary Intervention to a Hypoplastic Reimplanted Left Main Coronary in an Adolescent

Nitin Chandra Mohan, BMBS,[a] Alexander Carpenter, MBChB, BSc,[b] Stephanie Connaire, MBBCh,[b] Serban Stoica, MBBS, MD,[a] Stephen Dorman, BM BCh, MA,[b] Thomas W. Johnson, BSc, MBBS, MD[a]

ABSTRACT

Critical ostial stenosis following reimplantation of an anomalous left main coronary artery is extremely rare. Currently, there is no consensus on management following diagnosis. This report demonstrates the feasibility of percutaneous coronary intervention in an adolescent with such a condition and emphasizes the importance of periprocedural multimodality imaging. (JACC Case Rep. 2024;29:102509) © 2024 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

CASE PRESENTATION

A 14-year-old boy sustained a cardiac arrest while mountain biking. He had 5 minutes of cardiopulmonary resuscitation and 1 shock delivered by an automated external defibrillator. From collateral history, the patient had reported exertional chest pain and presyncopal symptoms 2 weeks before presentation. There was no past medical history or any family history of cardiac disease/sudden death. On arrival at the local hospital, the patient was hemodynamically stable but restless, with decorticate posturing; therefore, he was intubated and ventilated for neuroprotective measures. Initial electrocardiogram (ECG); biochemistry; and trauma head, chest, and abdomen computed tomography (CT) scan did not identify any overt acute pathologies. Further to this, the patient underwent a transthoracic echocardiogram (TTE) and CT of the coronary arteries (CTCA) (**Figure 1A**), both of which were suggestive of an intramural and interarterial left main coronary artery that originated from the right coronary cusp. A decision was made to treat surgically. Following sternotomy, the anomalous left main coronary artery (LMCA) had an interarterial course with a juxtacommissural ostium but was not intramural as suggested on noninvasive imaging. Furthermore, the native proximal LMCA was

LEARNING OBJECTIVES

- To understand how available adjuncts and multimodality imaging could be used to overcome the challenges of PCI in an adolescent with ostial stenosis after reimplantation surgery.
- To recognize that adolescents who have undergone PCI will require close follow-up because the stented artery will grow in size, potentially requiring further future intervention.

From the [a]Translational Health Sciences Department, University of Bristol, Bristol, United Kingdom; and the [b]Cardiology Department, University Hospitals Bristol and Weston NHS Trust, Bristol, United Kingdom.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received May 9, 2024; accepted May 23, 2024.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2024.102509

ABBREVIATIONS AND ACRONYMS

CT = computed tomography

CTCA = computed tomography of coronary arteries

DES = drug-eluting stent

ECG = electrocardiogram

IVUS = intravascular ultrasound

LMCA = left main coronary artery

PCI = percutaneous coronary intervention

TTE = transthoracic echocardiogram

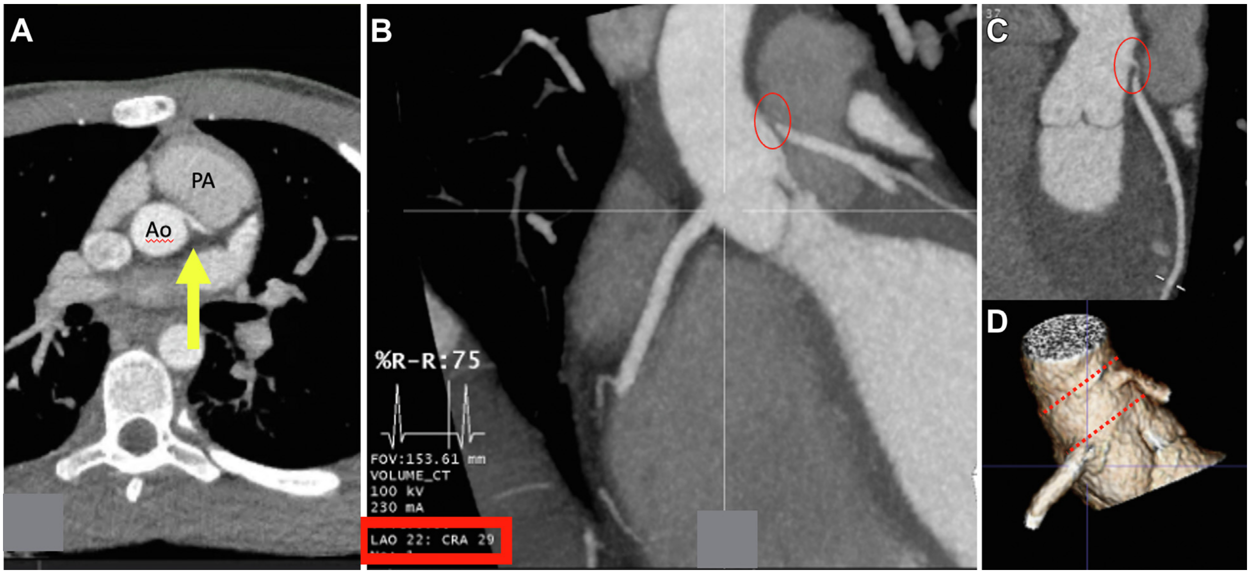
hypoplastic and had to be reimplanted more anteriorly than usual because the left ventricle was initially ischemic when coming off cardiopulmonary bypass.

Three months postoperatively, the patient underwent an exercise stress ECG and TTE. During peak exercise, there was evidence of transient ST-segment elevation in avR on ECG and anterior wall hypokinesia on TTE. A repeat CTCA was performed, which showed critical stenosis of the newly implanted LMCA (Figures 1B and 1C). Following a combined adult and pediatric meeting, a decision was made in favor of percutaneous coronary intervention (PCI) of the LMCA stenosis.

PCI was performed under general anesthesia at our high-volume regional cardiac center. Given the anatomy of the aortic root and size of the coronary artery, the procedure was elected to be performed by one of our senior adult interventional cardiologists. Because the experience operating in such patients was limited, the procedure was performed with close collaboration with adult and pediatric cardiologists with congenital cardiothoracic surgery backup onsite.

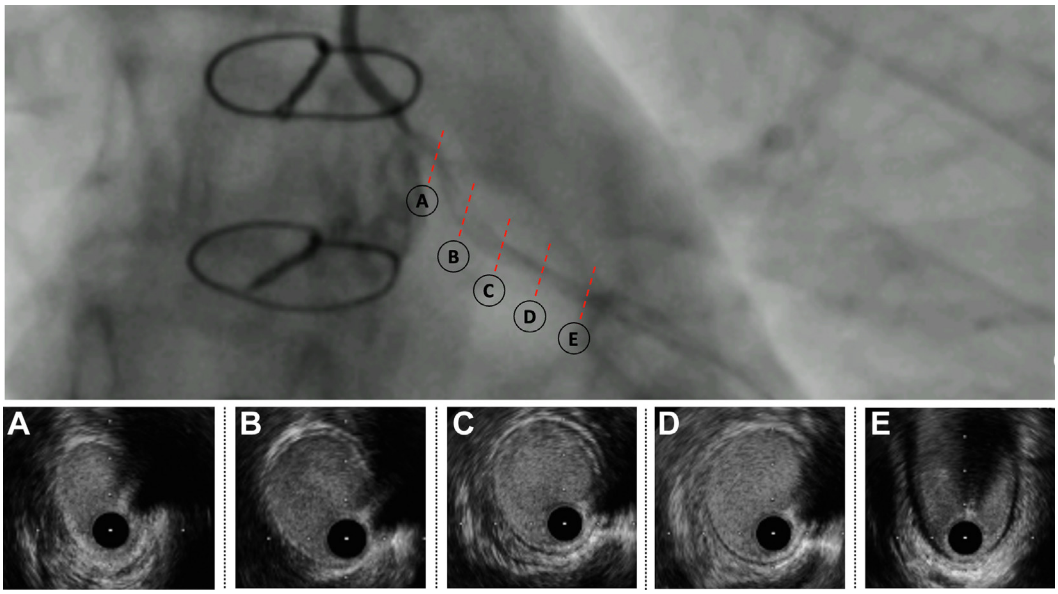
The LMCA was difficult to identify angiographically because its reinsertion point was high and anterior (Figures 1D and 2). This challenging anatomy, associated with a smaller aortic dimension than encountered in adult intervention, prevented engagement of the left main ostium through the right radial artery or right femoral artery, despite attempts with multiple guiding catheters. Periprocedural evaluation of the CTCA with determination of the ideal angiographic projection to delineate the LMCA ostium, combined with a left radial artery approach using a customized reshaped 5-F Judkins right guide catheter, using steam heating, ultimately facilitated engagement of the LMCA ostium. A HI-TORQUE WHISPER LS guidewire (Abbott Vascular) was passed into the distal left anterior descending artery, followed by a Sion (Asahi Intecc Co) blue wire to the left circumflex artery for additional support. Intravascular ultrasound (IVUS) evaluation confirmed an elliptical LMCA with reduced lumen area of 4.0 to 4.5 mm^2 (Figures 2A to 2E). The stenotic segment was predilated with a 3.0-mm noncompliant balloon and a 3.0-mm cutting balloon. Repeat IVUS examination confirmed reasonable modification, and therefore we proceeded with deploying a 3.5 × 16-mm Synergy Megatron (Boston Scientific) drug-eluting stent (DES). The stent was purposefully oversized to facilitate future postdilatation with growth of the patient. The DES was postdilated with a 3.5-mm noncompliant balloon to 14 atm, which resulted in a minimum stent

FIGURE 1 CT Coronary Angiography Pre- and Post-Cardiac Surgery



(A) The left main coronary artery (LMCA) (yellow arrow) arising from the right coronary cusp with an interarterial course demonstrated on axial computed tomography (CT). (B, C) Postoperative ostial LMCA stenosis (red circle) on reconstructed CT with optimal angiographic orientation (red box) for catheter engagement. (D) A superior and anterior placement of the LMCA in comparison to the right coronary artery. Ao = aorta; PA = pulmonary artery.

FIGURE 2 Fluoroscopic Frame Demonstrating Critical Stenosis of the Reimplanted Left Main Coronary Artery With Corresponding Intravascular Ultrasound Frames



Frame A of the intravascular ultrasound also demonstrates a "slit-like" left main coronary artery ostium. See Videos 1 to 4.

area in the LMCA of 7.5 mm^2 on IVUS. Further post-dilatation was performed, which increased the minimum stent area to 8.5 mm^2 (Figure 3A) on final IVUS. The end angiographic result revealed TIMI flow grade 3 to an unobstructed stented LMCA with no immediate complications (Figure 3A).

Two months postoperatively, the patient has returned to cycling. He is free of chest pain or presyncopal symptoms and remains under follow-up. Follow-up CTCA performed 4 months postoperatively revealed a patent, nondeformed stent (Figures 3B and 3C).

QUESTION 1: WHAT IS THE MANAGEMENT FOR PATIENTS WITH ANOMALOUS CORONARY ARTERIES ARISING FROM THE OPPOSITE SINUS OF VALSALVA?

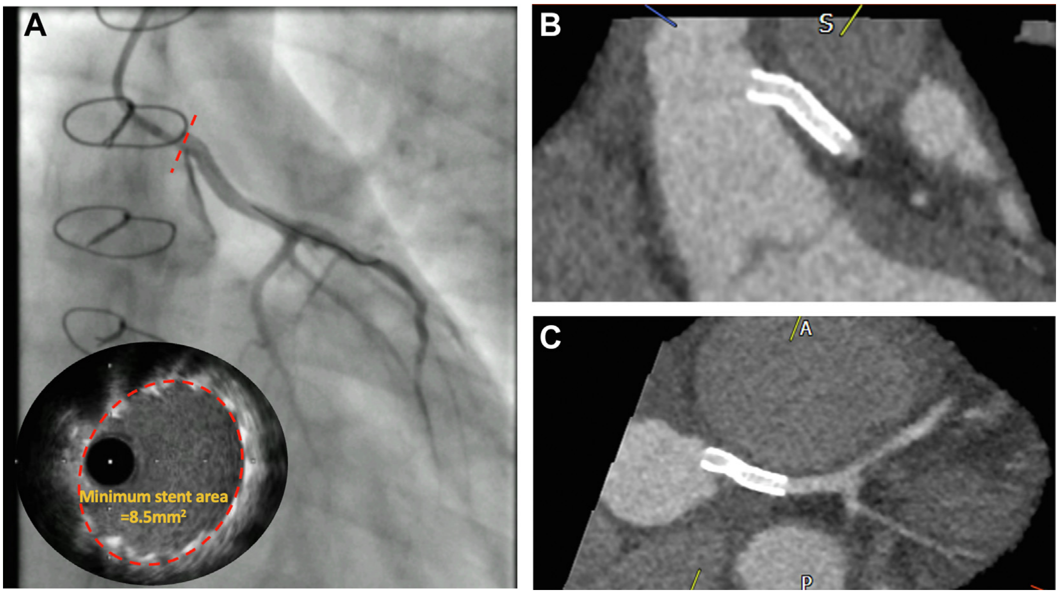
Anomalous coronary arteries arising from the opposite sinus of Valsalva are associated with an increased risk of sudden death, particularly when the artery takes an interarterial course.[1] At present, intervention is advised only in cases of high-risk coronary anatomy (slit-like orifice, acute-angle takeoff, orifice of >1 cm above the sinotubular junction), angina, or inducible ischemia in the relevant coronary territory. According to the European Society of Cardiology guidelines, CTCA is the preferred first-line modality to evaluate for high-risk anatomy.[2] For cases of anomalous coronary arteries with separate origins, complete coronary reimplantation may be favored. Like an arterial switch procedure, a coronary button is excised and reattached to the correct aortic sinus.[3]

QUESTION 2: WHAT IS THE INCIDENCE AND MECHANISM OF OSTIAL CORONARY STENOSIS FOLLOWING REIMPLANTATION SURGERY FOR ANOMALOUS CORONARY ARTERIES ARISING FROM THE OPPOSITE SINUS OF VALSALVA?

Although it is difficult to give estimates of postoperative coronary complications, the original Bentall procedure has been associated with a 6% incidence of coronary ostial stenosis following coronary button reimplantation.[1]

Several mechanisms behind coronary stenosis postoperatively have been described. These include damage during instrumentation, local pressure necrosis and subsequent intimal proliferation, postoperative immune reactions, and genetic predisposition.[4] In our case, reimplantation of the LMCA in its usual anatomic position was not well tolerated by the patient intraoperatively,

FIGURE 3 **Final Coronary Angiography Post-Intervention and Follow-Up CT Scan**



(A) Final angiographic run illustrating an unimpinged ostial left main segment. Embedded is a final intravascular ultrasound frame of the left main ostium demonstrating acceptable minimal stent area and absence of the "slit-like" appearance. (B, C) A patent nondeformed stent in the left main coronary artery. See Videos 5 and 6.

necessitating a more anterior placement. Although the exact mechanism of stenosis is unclear, it is likely that the hypoplastic LMCA was a major contributing factor.

QUESTION 3: WHAT IS THE MANAGEMENT OF PATIENTS WHO DEVELOP CRITICAL CORONARY STENOSIS AFTER REIMPLANTATION SURGERY?

Currently, there is a lack of consensus on managing this condition. Data on long-term outcomes of bypass grafting in this cohort are missing. Additionally, our multidisciplinary heart team believed that the risks of repeat open heart surgery could be avoided. On balance, PCI was the opted strategy because it carried fewer procedural risks than repeat surgery, and its feasibility has been demonstrated in adolescents.

QUESTION 4: WHAT ARE THE CONSIDERATIONS FOR PCI OF A STENOSED REIMPLANTED CORONARY OSTIUM?

PCI in this setting remains relatively novel and should be undertaken by highly experienced operators with collaboration between the pediatric and adult interventional cardiologists during the procedure. We must be mindful of the elastic recoil and cognizant that the stented artery will increase in size, requiring further postdilatation in the future. It is therefore important to choose a DES with high radial strength and a good expansion profile and to closely monitor these patients. From a technical perspective, engaging the reimplanted coronary with standard preshaped catheters may not be straightforward. Alternate access sites should be considered for better guide support. To add to this, reshaping catheters using steam or boiling water may be required for coaxial engagement. Although CTCA before PCI is not part of our routine practice, we find it highly valuable in patients who have undergone prior surgery. In this case, it allowed us to evaluate for postsurgical complications before subjecting the patient to an invasive and relatively higher-risk procedure. Having the information beforehand also gave us the opportunity to preplan our intervention and assist with the procedure by establishing the optimal x-ray film orientation (**Figure 1B**) for guide catheter engagement. Procedural success also relied on the use of intracoronary imaging, which allowed identification of the etiology of coronary stenosis and guide pre- and post-PCI strategy.

QUESTION 5: HOW DO WE MONITOR PATIENTS WHO HAVE UNDERGONE PCI OF A REIMPLANTED CORONARY BUTTON?

Currently, there are no recommendations around monitoring adolescents who have undergone PCI. The left main coronary artery diameter has been shown to increase 4-fold from 1 mm to 4.5 mm between infancy and 17 years of age.[5] Previous reports of PCI in the pediatric population (primarily involving infants) have demonstrated the need for postdilation in at-least three-quarters of cases.[6,7] Our patient underwent a follow-up CTCA 4 months postprocedurally and will be clinically assessed on an annual basis. Interim cross-sectional imaging will be considered if there any concerns of ongoing ischemia. Otherwise, a repeat CTCA will be performed when the patient is 17 years old, 2.5 years after index procedure. If there is a significant increase in vessel caliber, we will consider postdilation following a heart team discussion.

CONCLUSIONS

To our knowledge, this is the first case of PCI to a reimplanted anomalous and hypoplastic LMCA. Multimodality imaging is strongly recommended to guide PCI in these patients, and close follow-up is mandated.

ACKNOWLEDGMENT Written consent has been obtained from the patient's next of kin. (The patient is 14 years old.)

FUNDING SUPPORT AND AUTHOR DISCLOSURES

Dr Johnson has received consultancy and speaker fees from Abbott Vascular, Boston Scientific, and Terumo and research grants from Abbott Vascular. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Thomas W. Johnson, Level 7, Queen's Building, Bristol Royal Infirmary, Bristol, BS2 8HW, United Kingdom. E-mail: tom.johnson@bristol.ac.uk.

REFERENCES

1. Gentile F, Castiglione V, De Caterina R. Coronary artery anomalies. *Circulation*. 2021;144(12):983–996.

2. Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC guidelines for the management of adult congenital heart disease. *Eur Heart J*. 2021;42(6):563–645.

3. Gaillard M, Pontailler M, Danial P, et al. Anomalous aortic origin of coronary arteries: an alternative to the unroofing strategy. *Eur J Cardiothorac Surg*. 2020;58(5):975–982.

4. Bekke K, Dadnegard H, Sigvardsen PE, Smerup M. Myocardial ischaemia caused by bilateral coronary ostial stenosis from pseudointimal membranes in a full root freestyle valve: a case report. *Eur Heart J Case Rep*. 2020;4(5):1–8.

5. Oberhoffer R, Lang D, Feilen K. The diameter of coronary arteries in infants and children without heart disease. *Eur J Pediatr*. 1989;148(5):389–392.

6. Chaszczewski KJ, Nicholson GT, Shahanavaz S, et al. Stent angioplasty for post-operative coronary artery stenosis in infants. *World J Pediatr Congenit Heart Surg*. 2022;13(2):203–207.

7. Stanfill R, Nykanen D, Osorio S, Whalen R, Burke RP, Zahn EM. Stent implantation is effective treatment of vascular stenosis in young infants with congenital heart disease: acute implantation and long-term follow-up results. *Catheter Cardiovasc Interv*. 2008;71(6):831–841.

KEY WORDS anomalous coronary artery, cardiac arrest in the young, percutaneous coronary intervention in an adolescent

APPENDIX For supplemental videos, please see the online version of this paper.