Received: 20 July 2023 | Revised: 2 October 2023 | Accepted: 3 October 2023

DOI: 10.1002/ccr3.8052

CASE REPORT

Clinical Case Reports Open Access WILEY

Percutaneous coronary intervention for ventricular fibrillation in the setting of an anomalous right coronary artery

Danish Saleh[1] | **Eric P. Cantey**[1] | **Emily P. Marogi**[2] | **Benjamin H. Freed**[1] | **Bradley P. Knight**[1] | **Roger A. de Freitas**[1] | **Ranya N. Sweis**[1] | **James D. Flaherty**[1]

[1]Department of Medicine, Division of Cardiology, Northwestern University Feinberg School of Medicine, Chicago, Illinois, USA

[2]Department of Medicine, University of California San Francisco School of Medicine, San Francisco, California, USA

Correspondence
Danish Saleh, Department of Medicine, Division of Cardiology, Feinberg School of Medicine at Northwestern University, 676 North St. Clair Street, Arkes Suite 600, Chicago, IL 60611, USA.
Email: danish.saleh@nm.org

Key Clinical Message

We present a case of a quadriplegic male who developed ventricular fibrillation associated with an anomalous aortic origin of the right coronary artery. Successful revascularization was achieved with percutaneous coronary intervention. This case highlights the application of an unconventional approach to resolve ischemia in a patient with prohibitive surgical risk.

KEYWORDS

anomalous coronary artery, anomalous right coronary artery, ischemia, percutaneous coronary intervention, ventricular fibrillation

1 | HISTORY OF PRESENTATION

Our patient is a 29-year-old bedbound male with quadriplegia related to injury at the fourth cervical vertebra (C4) and a history of atrial fibrillation, who was found to be critically ill with a urinary tract infection, prompting hospital admission. On the fifth day, the patient developed cardiac arrest with ventricular fibrillation. Return of spontaneous circulation (ROSC) was achieved after defibrillation. Patient was supported with electrolyte repletion, anti-arrhythmic therapy, and transvenous pacemaker placement with rapid-ventricular pacing. Targeted-temperature management was enacted per protocol.

2 | PHYSICAL EXAMINATION

On admission, cardiovascular examination was without tachycardia and/or obvious murmurs though the rhythm appeared irregular. Patient had shallow breaths and poor inspiratory effort. Abdomen was diffusely tender, and lower extremities contracted.

3 | PAST MEDICAL HISTORY

Medical history is notable for C4 quadriplegia secondary to a motor-vehicle accident, autonomic-dysfunction, neurogenic-bladder, decubitus ulcers, and recurrent urinary tract

and bone infections (left ischial tuberosity osteomyelitis) requiring intravenous antibiotics. Patient had atrial fibrillation which was previously managed with propafenone and metoprolol. These medications were not well-tolerated and were unilaterally discontinued by the patient a month prior to admission.

4 | DIFFERENTIAL DIAGNOSIS

The differential diagnosis for this patient's ventricular fibrillation included myocardial ischemia, structural heart disease including myocardial scar and/or severe pump dysfunction, congenital and acquired QT-prolonging conditions, severe electrolyte disturbances especially hypokalemia and/or hypomagnesemia, and hemodynamic or metabolic stress in the setting of acute illness.

5 | PERTINENT DATA

At the time of arrest, laboratories were notable for potassium of 3.0 mEq/L with hemoglobin at baseline (11.7 g/dL). Tele-strips at the time of arrest demonstrated R-on-T phenomenon with ventricular fibrillation (Figure 1). On Admission, high-sensitivity troponin was 12 and initial electrocardiogram (EKG) showed atrial fibrillation alternating with sinus rhythm and premature ventricular contractions with a corrected QT interval of 431 ms (Figure 2A). On admission, two-dimensional transthoracic echocardiography was notable for mildly decreased left ventricular systolic function, and an ejection fraction of 50% with beat-to-beat variation due to frequent ectopy.

6 | INVESTIGATIONS AND MANAGEMENT

Post-arrest EKG was notable for ventricular quadrigeminy and corrected QT interval of 554 ms for sinus beats (Figure 2B) without clear evidence of ischemia. Echocardiogram revealed a depressed ejection fraction of 21% in the immediate post-arrest period which improved to 40% within 1 week. Cardiac magnetic resonance imaging (MRI) demonstrated a moderately reduced ejection fraction of 40% without evidence of myocardial fibrosis. Coronary computed tomography (CT) scan angiogram revealed an anomalous right coronary artery (aRCA), emerging from the left main, with significant narrowing between the aortic and pulmonary trunks (Figure 3A,B). Calculation of fractional flow reserve (FFR) of the RCA demonstrated a hemodynamically significant anatomic lesion (FFR = 0.51; Figure 3C). Coronary angiogram confirmed significant narrowing of aRCA emerging from the left main (Figure 4A). Intravascular ultrasound (IVUS) demonstrated a proximal aRCA with extrinsic compression by aortic and pulmonary trunks and ellipse-like cross-section with minimal luminal area of 3.4 mm^2. Distally, the aRCA had a luminal area of 8.7 mm^2 (Figure 4B,C). The patient underwent PCI to the ostial and proximal RCA with a Xience™ 3.25 × 28 mm drug-eluting stent. Post-intervention, IVUS demonstrated rectification of extrinsic compression and recovery of luminal cross-section area (7 mm^2) associating with TIMI 3 flow on angiogram (Figure 5A,B). No arrhythmias or complications were observed during the procedure. Post-intervention, the patient was managed with dual-antiplatelet therapy (DAPT) and anticoagulation (for atrial fibrillation and peripherally-inserted central catheter associated thrombus). Prior to discharge, the patient was transitioned to oral amiodarone and an implantable cardioverter defibrillator (ICD) was placed for secondary prevention. Possible osteomyelitis was managed with continued antibiotics.

7 | FOLLOW-UP

At 18-months follow-up, there were no further ventricular arrhythmias and/or ICD discharges observed. Moreover, no bleeding complications were reported.

8 | DISCUSSION

Anomalous aortic origin of the left coronary artery (AAOLCA) or right coronary artery (AAORCA) have independently been linked to sudden cardiac death in young individuals.[1–3] Myocardial ischemia is thought to occur during periods of stress, such as physical activity or periods of heightened metabolic demand, as increased myocardial demand cannot be met due to anatomic obstruction, including vessel compression by adjacent structures (i.e., aorta and pulmonary trunk), acute-angle take-off, and/or intramural coronary penetration.[4] Nevertheless, these lesions are observed infrequently and have only been reported in 0.1–1% of births.[5]

The optimal approach to ensure durable myocardial perfusion in the setting of a clinical and/or a hemodynamically significant anomalous coronary artery lesion remains unclear. The 2018 AHA/ACC guidelines for the management of congenital heart disease in adults recommend surgery (Class I, Level B-NR) for any AAOCA with myocardial ischemia or AAOLCA without

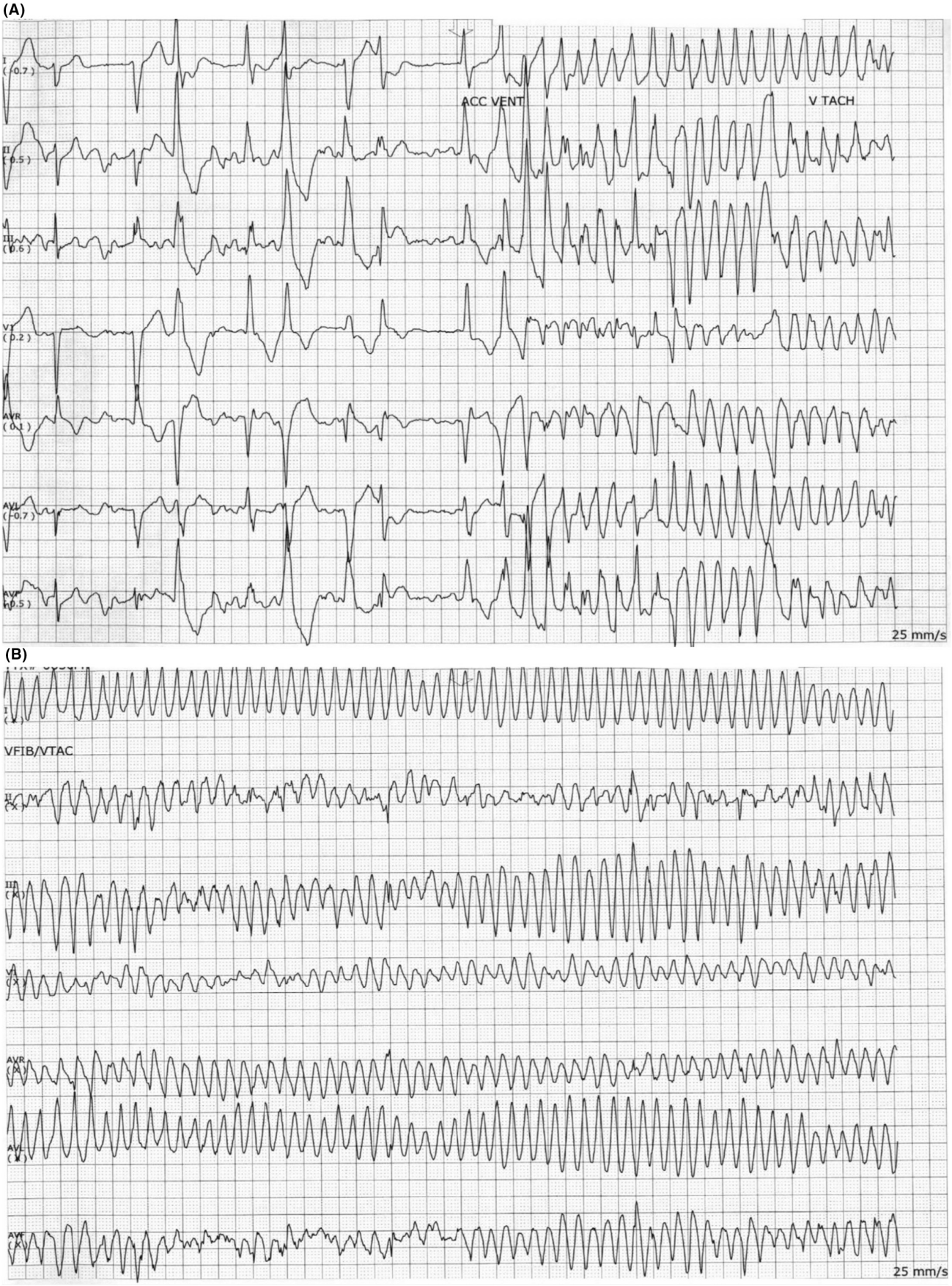

FIGURE 1 (A) A six-lead telemetry rhythm strip showing atrial fibrillation with natively conducted QRS complexes, aberrantly conducted QRS complexes, PVCs, and the onset of ventricular fibrillation. (B) Telemetry strip demonstrating ventricular fibrillation.

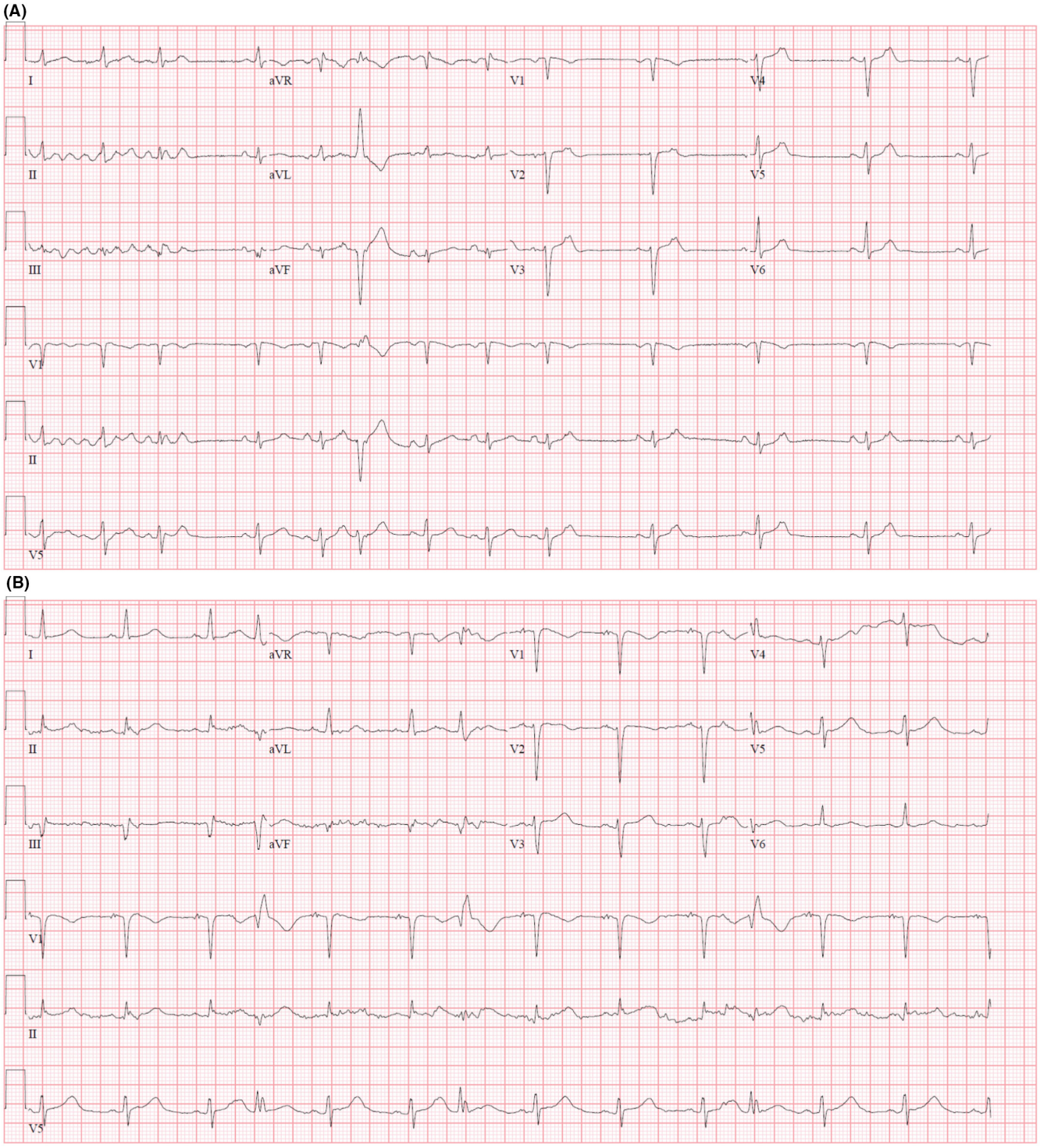

FIGURE 2 (A) Admission EKG, QTc ~431. (B) Post-arrest EKG, QTc ~554.

evidence of ischemia (Class IIa/Level C-LD).[6] Similarly, The American Association for Thoracic Surgeons (AATS) recommend surgery (Class I/Level B) in patients with asymptomatic or symptomatic AAOLCA or symptomatic AAORCA.[7] Surgical approaches include coronary "unroofing," coronary re-implantation with and/or neo-orifice generation in the correct coronary sinus, pulmonary artery translocation, and coronary-artery bypass grafting (CABG). The AATS advises consideration of PCI for adults with high surgical risk (Class IIb/Level C).[7]

In patients with AAORCA, PCI has been shown to be an effective and low-risk alternative to surgery in limited long-term studies.[8,9] PCI-associated remodeling

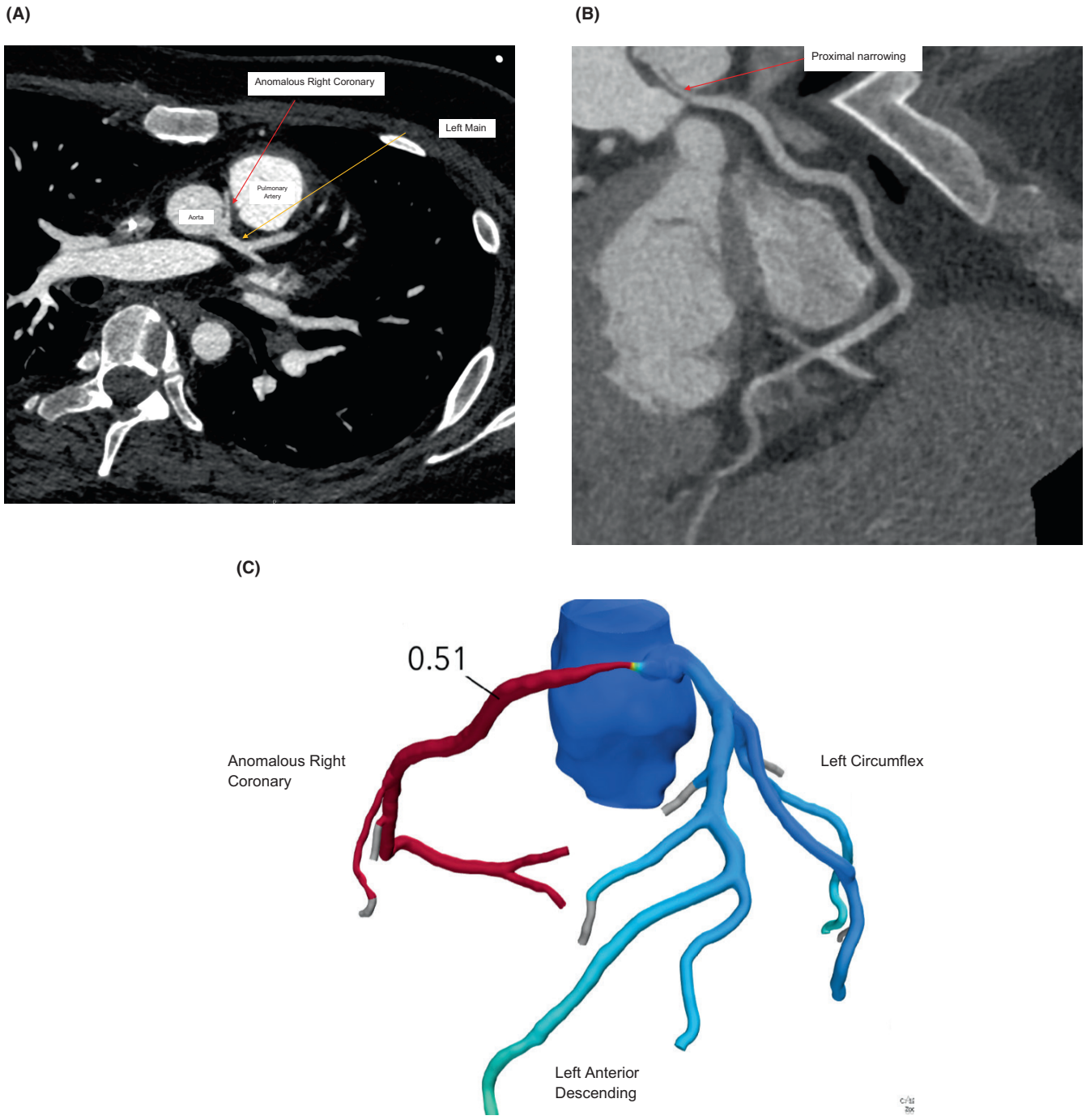

FIGURE 3 (A) CT-coronary angiogram demonstrating emergence of an aRCA from the left main. (B) Dedicated RCA view. (C) 3D graphical reconstruction of coronary anatomy with FFR quantitation of coronary flow across proximal aRCA stenosis. FFR of RCA ~0.51.

increases coronary lumen area and features resolution of ischemia.[10] Historically, surgical approaches have been preferred to percutaneous approaches in pediatric patients because of anticipated cardiac/coronary growth and concern for long-term stent-associated complications and/or durability. In our patient, who was a poor candidate for post-operative rehabilitation, a minimally invasive approach with PCI offered a desired alternative when compared to surgical revascularization to mitigate myocardial ischemia. Moreover, it is well-recognized that PCI and the need for DAPT therapy can increase bleeding risk; however, our patient was tolerating anticoagulation for atrial fibrillation and did not have underlying diatheses and/or history of bleeding. Indeed, no bleeding complications were appreciated in the post-procedural period. Of note, a retrospective study of patients with paraplegia

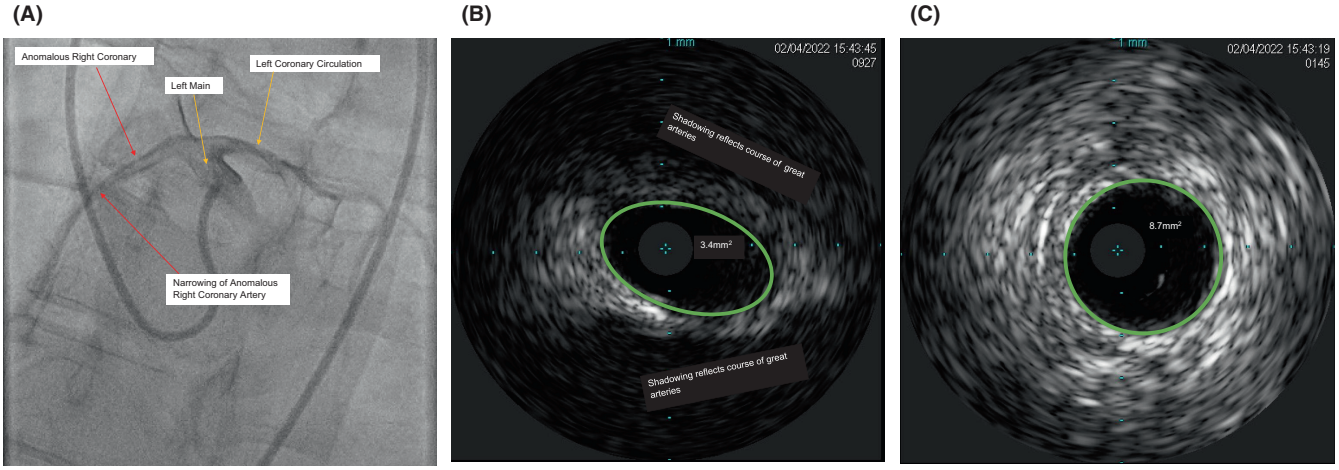

FIGURE 4 (A) Pre-intervention coronary angiogram demonstrating aRCA emerging from the Left Main and region of narrowing. (B) Pre-intervention IVUS of proximal aRCA demonstrating ellipse-like luminal compression with luminal area of 3.4 mm^2. Shadowing reflects anatomy of the aortic and pulmonary trunks. (C) Pre-intervention IVUS of the aRCA distal to proximal narrowing with luminal area of 8.7 mm^2.

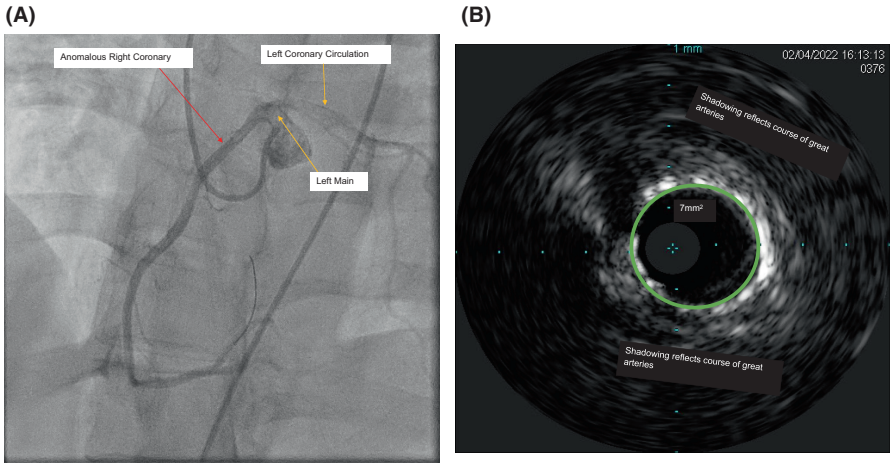

FIGURE 5 (A) Post-intervention coronary angiogram demonstrating left main, left coronary circulation, and aRCA with resolution of proximal narrowing. (B) Post-intervention IVUS of proximal aRCA post PCI with rectification of luminal integrity and recovery of luminal area (7 mm^2). Shadowing reflects anatomy of the aortic and pulmonary trunks.

and myocardial infarction who underwent revascularization with PCI had lower in-hospital mortality compared to those who underwent coronary artery bypass grafting.[7] While the safety and durability of PCI in the pediatric population has yet to be examined, emerging data support consideration of PCI in adults with AAOCA and high surgical risk.[8–11]

9 | CONCLUSION

We suspect that ventricular arrhythmia was the manifestation of ischemia in the setting of congenital flow-limiting stenosis within the right coronary artery. The ventricular fibrillation threshold was likely lowered by a hypermetabolic state of sepsis, hypotension, and underlying electrolyte abnormalities (hypokalemia). In this young adult with quadriplegia who was a poor candidate for post-surgical rehabilitation, a minimally invasive approach with PCI offered a solution to maintain coronary patency, mitigate myocardial ischemia, and lower the risk of recurrent cardiac arrest. Although PCI is not a first-line approach for revascularization in the setting of anomalous coronary anatomy, the solution employed in this case is supported by clinical guidance for patients at high operable risk.[8]

AUTHOR CONTRIBUTIONS

Danish Saleh: Conceptualization; data curation; methodology; project administration; writing – original draft; writing – review and editing. **Eric P. Cantey:** Data curation; writing – review and editing. **Emily P. Marogi:** Writing – original draft. **Benjamin H. Freed:** Writing – review and editing. **Bradley P. Knight:** Writing – review and editing. **Roger A. de Freitas:** Writing – review and editing. **Ranya N. Sweis:** Writing – review and editing. **James D. Flaherty:** Conceptualization; supervision; writing – review and editing.

ACKNOWLEDGMENT
None.

FUNDING INFORMATION
None.

CONFLICT OF INTEREST STATEMENT
Dr. Knight has received research grants from and has served as a consultant for Abbott, Biotronik, Boston Scientific, and Medtronic. Dr. Sweis is a part of the Speakers' Bureau with Edwards Lifesciences.

DATA AVAILABILITY STATEMENT
Data sharing is not applicable to this article as no new data were created or analyzed in this study.

ETHICS STATEMENT
We confirm that this manuscript has not been published elsewhere and is not under consideration by another journal. We have removed all identifiable information from this case study for academic and educational purposes. Accordingly, this body of work does not constitute "research," and institutional review board (IRB) approval was not required or obtained. All authors have approved the manuscript and agree with submission to *Clinical Case Reports*.

CONSENT
Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

ORCID
Danish Saleh https://orcid.org/0000-0001-8562-7539

REFERENCES
1. Angelini P. Coronary artery anomalies: an entity in search of identity. *Circulation*. 2007;115:1296-1305.
2. Cheitlin MD, De Castro M, McAllister HA. Sudden death as a complication of anomalous left coronary origin from the anterior sinus of Valsalva, a not-so-minor congenital anomaly. *Circulation*. 1974;50:780-787.
3. Barth CW III, Roberts WC. Left main coronary artery originating from the right sinus of Valsalva and coursing between aorta and pulmonary trunk. *JACC*. 1986;7:373.
4. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *JACC*. 1992;20:640-647.
5. Brothers JA, Frommelt MA, Jaquiss RDB, Myerburg RJ, Fraser CD Jr, Tweddell JS. Expert consensus guidelines: anomalous aortic origin of a coronary artery. *J Thorac Cardiovasc Surg*. 2017;153:1440-1457.
6. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American college of cardiology/American heart association task force on clinical practice guidelines. *J Am Coll Cardiol*. 2019;73:1494-1563.
7. Lu SF, Lu LX, Smith SC Jr, Dai X. Acute myocardial infarction in patients with paraplegia: characteristics, management, and outcomes. *Am J Med*. 2017;131:574.e1-574.e11.
8. Angelini P, Walmsley RP, Libreros A, Ott DA. Symptomatic anomalous origination of the left coronary artery from the opposite sinus of valsalva. Clinical presentations, diagnosis, and surgical repair. *Tex Heart Inst J*. 2006;33:171-179.
9. Darki A, Motiwala A, Bahos L, et al. Technical success and long-term outcomes after anomalous right coronary artery stenting with cardiac computed tomography angiography correlation. *Catheter Cardiovasc Interv*. 2020;96:320-327.
10. Angelini P, Uribe C, Monge J, Tobis JM, Elayda MA, Willerson JT. Origin of the right coronary artery from the opposite sinus of valsalva in adults: characterization by intravascular ultrasonography at baseline and after stent angioplasty. *Catheter Cardiovasc Interv*. 2015;86:199-208.
11. Aubry P, du Fretay XH, Boudvillain O, Degrell P, ANOCOR Working Group. Place of angioplasty for coronary artery anomalies with Interarterial course. *Front Cardiovasc Med*. 2021;7.

How to cite this article: Saleh D, Cantey EP, Marogi EP, et al. Percutaneous coronary intervention for ventricular fibrillation in the setting of an anomalous right coronary artery. *Clin Case Rep*. 2023;11:e8052. doi:10.1002/ccr3.8052