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EDITORIAL COMMENT

The Difficulty in Navigating a Storm Without a Compass

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A common theme in congenital cardiology is the fact that patients are living longer with improved contemporary management and surgical strategies,[1] which leads inevitably to uncharted waters regarding long-term surveillance and management for these patients. In certain forms of pediatric and congenital heart disease, there has been the ability to collect outcomes data and formulate management recommendations and guidelines, such as in patients with tetralogy of Fallot (ToF) or hypertrophic cardiomyopathy (HCM).[2-6] However, no such guidelines exist for patients with the rarer anomalous left coronary artery from the pulmonary artery (ALCAPA), and the case report from Jeong et al[7] in this issue of *JACC: Case Reports* demonstrates the uncertainty in which pediatric and congenital practitioners often find ourselves navigating when it comes to patient care.

Like in patients with ToF and HCM, a major concern with ALCAPA lies in the risk for sudden cardiac death (SCD) due to the impact of the disease on ventricular scarring and substrate for malignant arrhythmias.[8,9] Investigation has identified multiple SCD risk factors in ToF and HCM, allowing for risk stratification and guideline-directed management.[2-6] There has been substantially less investigation into ALCAPA outcomes, and even within these reports, the rhythm data are extremely scarce.[10-13] The potential for proarrhythmic effect due to preoperative abnormal coronary perfusion and postoperative ventricular scarring makes mechanistic sense and must be a focus of surveillance as patients age.

Although there are a handful of case reports of ALCAPA diagnoses being revealed by presentations with malignant arrhythmias in adults,[14,15] the Jeong et al[7] report demonstrates the arrhythmia risk that patients with repaired ALCAPA inherit after their definitive surgical repair. In this particular case, the patient's acute presentation and management trajectory followed the appropriate flow through established ventricular tachycardia (VT) management algorithms for patients with underlying structural heart disease,[16] culminating in extensive diagnostic workup and ultimate secondary prevention implantable cardioverter-defibrillator (ICD) implantation and VT ablation. Although this patient followed an algorithmic progression through established guidelines, the investigators highlight the potential for missed diagnosis and risk assessment before his acute presentation.

The patient underwent regular ambulatory rhythm monitoring and echocardiography, with no identified arrhythmias nor ventricular systolic dysfunction. However, in the year leading up to the acute presentation, he underwent multiple evaluations for symptoms that in hindsight may have reflected earlier presentations of malignant arrhythmias. Should he have undergone screening with cardiac magnetic resonance imaging (CMR) earlier based on his previous history of known scar? How reliable was his ventricular function assessment by echo in comparison to CMR? Should he have undergone more aggressive and formal electrophysiological evaluation with ventricular stimulation testing and/or loop recorder implantation rather than just regular Holter monitoring? And if there had been demonstrated clinical or inducible ventricular arrhythmias, what would have been the appropriate management strategy? Sentencing a pediatric patient to a lifetime of amiodarone or ICD implantation is not a decision that is taken lightly. Secondary prevention ICD

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implantation is one thing, but a primary prevention decision is entirely different. With discrepant left ventricular function determinations by echo and CMR, this may have limited antiarrhythmic medication options even further. The mechanistic overlap between the ALCAPA disease substrate and post-myocardial infarction substrate would seem to allow for reasonable use of post-myocardial infarction management algorithms to guide management of ALCAPA patients. However, the patient populations are quite different, so is this really an appropriate use of guidelines?

As is a typical refrain in the congenital world, there simply needs to be more data. Although the ALCAPA patient population is not nearly as large as that of non-congenital adults with coronary disease (nor ToF or HCM patient populations), investigation of patient outcomes and risk of SCD is just as important and warrants focus to attempt to provide a management framework for providers. Existing datasets should be mined for data reflecting scar and ventricular function on CMR, baseline electrocardiogram data, Holter data, and incidence of malignant arrhythmias and SCD. As is often required in the pediatric and congenital world with uncommon diseases, multicenter studies should be pursued to pool together contemporary postoperative outcomes and give more power to the conclusions that are drawn. Until these things are done, this report is a reminder of the murky reality that patients with congenital heart disease and their providers will continue to operate in.

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