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Editorial

Predicting acquired cardiovascular disease in adults with congenital heart disease is risky business

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Acquired cardiovascular disease (ASCVD) remains the leading cause of death in the United States. Over two decades ago, myocardial infarction became the primary contributing cause of death for adults with non-cyanotic congenital heart disease [1]. Over 80% of adults with congenital heart disease (CHD) have been identified to have one or more acquired cardiovascular risk factors [2]. Additionally, certain types of CHD and/or prior surgical repair have been associated with an increased risk of ASCVD [3].

In the current study, Hacker AL et al. use the Prospective Cardiovascular Munster (PROCAM) 10 year risk score for cardiovascular event to assess ASCVD risk in adults with CHD. The authors demonstrated that the absolute 10 year risk was lower in adults with CHD as compared to the general population (2.5 ± 4.9% in adults with CHD as compared to 4.8 ± 5.2% in the general population). These findings are similar to studies that have evaluated the ASCVD risk score in the ACHD population as compared to the general population [4]. This score is largely driven by tobacco use and prescription of antihypertensive medications, which were markedly different among the two groups. Abnormal lipids were infrequently seen in this population. Importantly, the Framingham Risk Assessment, Reynolds Risk Score, and ASCVD risk estimator were derived from an older population ages 40 to 79 years with no history of CHD. When these risk scores were calculated for the patients within the actual age range, the ASCVD risk was higher in one cohort suggesting that these risk scores may underestimate the risk of ASCVD in this younger population [5].

None of these risk calculators, including PROCAM, have been validated in the adult congenital heart disease (ACHD) population. Additionally, PROCAM has not been prospectively validated for ASCVD events in the general population and has been documented to overestimate risk in validation attempts [6]. Even though these risk scores are well established, some individuals with high scores do not develop ASCVD in the next 10 years while others with low scores demonstrate subclinical atherosclerosis and/or myocardial infarction [7]. It is imperative for these risk scores to be validated in prospective, outcomes-based cohorts of ACHD patients. These traditional risk scores may need to be adjusted to include ACHD specific risk factors and prospective study of the predictive ability of risk scores to determine outcomes better than usual clinical care must then be demonstrated.

The need for risk prediction for ASCVD events in the ACHD population is clear. In a cohort of individuals with CHD that underwent catheterization, 9% had evidence of coronary atherosclerosis [8]. This data may increasingly be actionable, as research has demonstrated a trend towards reduced survival in patients who underwent repeat CHD surgery with concomitant coronary artery bypass grafts [3]. With the rising rate of coronary cardiovascular events in the ACHD population, there are several important risk contributors which a relevant risk calculator must address.

Novel risk factors may include type of CHD or surgical repair, which may place some individuals with CHD at increased risk. Patients with CAD documented on cardiac catheterization are older with greater CHD complexity as well as hypertension (HTN) and hyperlipidemia [8]. CAD has also been implicated as a leading cause of death after repaired coarctation. Coarctation patients have subclinical atherosclerosis identifiable on computed tomography in high rates when compared to patients without coarctation [7] and demonstrate higher rates of HTN, heart failure and stroke [3]. Premature ASCVD in coarctation is likely multifactorial, influenced by the increased prevalence of HTN in addition to persistent endothelial dysfunction and impaired arterial reactivity from a primary vascular abnormality in coarctation [3]. Coronary anomalies and surgical coronary manipulation should also be considered in assessing relevant risk factors for the development of CAD and associated events. Coronary lesions have been seen in 5% of late survivors of transposition of the great arteries (TGA) after arterial switch operation (ASO) [9]. Coronary stenoses in TGA ASO are often asymptomatic and the new AHA/ACC guidelines have advocated routine evaluation with coronary angiography or computed tomography coronary angiography in asymptomatic TGA ASO patients [10]. Despite these findings, long-term outcome has been good in this population with few adverse coronary events. However, this population remains

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quite young and the impact of acquired coronary heart disease is unknown. Prospective study for risk assessment in this unique population is an important future direction for research.

The incorporation of novel risk factors that are unique to the ACHD population with traditional risk factors may offer better discrimination for future ASCVD. However, one of the key opportunities of risk calculators is whether it will change clinical management. Despite patients who met clinical guidelines for drug therapy such as a statin by ASCVD risk assessment, more than 50% of ACHD cases did not receive primary prevention statin [4]. As individuals with ACHD survive into adulthood, acquired comorbidities and traditional risk factors such as HTN, diabetes, dyslipidemia and smoking may begin to define their outcome by increasing morbidity and health care utilization. Screening for cardiovascular risk factors and modifying these risks earlier in life may affect their long-term outcome. Whereas much of this will happen as part of routine care, we are still dealing with a high rate of loss to follow up and an important issue of access to tertiary care.

Comprehensive risk modeling, including traditional ASCVD as well as unique CHD risk factors will enable our networks of care to identify higher risk patients in the community and designate efforts at enrollment into ACHD clinics and resource utilization to the populations who would benefit the most, while better controlling the rising rates of anxiety in those individuals with CHD who are stable and well. Continued study of risk prediction is essential. Researchers must be realistic in what drives their models and work with others to create actionable rather than siloed data as this population grows and ages.

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

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