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EDITORIAL COMMENT

Biking More With Congenital Heart Disease

Stratifying Risk in the New Generation of Adult Patients*

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Understanding midterm outcomes in congenital heart disease (CHD), particularly in young adults, suffers from several important areas of known uncertainty. The first, and maybe the most important, is that the overall mortality rates for the patients are low and that the mortality rates for interventions are low. Those terrific realities have become more apparent in the recent era.[1] Related to that is that the overall approach to congenital lesions has shifted. Infant and childhood aortic stenosis now starts with catheter-based interventions, d-transposition of the great vessels is treated with an arterial switch rather than an atrial switch, tetralogy of Fallot is repaired in infancy, often with very limited outflow tract incisions. Residual pulmonary insufficiency is aggressively managed in many settings based on numeric criteria for right ventricular dilation and not on symptoms. In each scenario data for what to do for patients in the 20s and 30s is invariably based on data obtained on patients who are now in their 40s to 60s.

The European medical care system, with its capacity to reliably capture both routine data and mortality, has resulted data sets that help to overcome these limits. SWEDCOM is a Swedish registry with extremely high enrollment of both children and adults who have echocardiographic diagnoses of CHD. Registry leadership invests energy in assuring its validity. The data in this study were captured in 2017 when there were 10,192 patients ≥19 years of age enrolled.[2]

In this issue of *JACC: Advances,* Wikner et al[3] critically evaluate the role of peak workload on overall mortality in a large cohort of adult CHD patients who underwent exercise testing. This is a relatively modern cohort with a median age of 27 years and with 75% of the cohort <41 years of age. This cohort has an overall mortality of 5.8% over a mean follow-up of 9.4 years; this which permits using mortality as the analytic endpoint. Exercise testing was done using a relatively simple bicycle ramp protocol with peak workload (alas not presented) and percent predicted workload as their primary outcome. The analysis focuses on those patients who have moderately (<70% and >50% predicted workload) to severely depressed (<50% predicted workload) workloads. That simple measure of strength and aerobic capacity is strongly associated with mortality with expected dose response. The hazard ratios are 2.3 and 5.6 for moderate and severely depressed workloads, respectively.

The research team does a careful job of looking at both anatomic, functional, and potentially behavioral risk factors. As we have come to expect systemic ventricular function, NYHA functional class and disease complexity combine to permit a reasonable prediction of outcome with an area under the receiver operator curve of ~0.8. The additive value of exercise testing was modest, though clearly positive. However, when modeling included both exercise

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capacity and NYHA functional classification, measured exercise capacity was a superior predictor.

There are a number of limitations which the authors address. In the SWEDCOM adult CHD cohort, only 37% of participants had exercise testing for analysis. Those who did not have exercise testing had a 1.9× higher mortality rate. The exercise cohort has heavily biased toward the moderate complexity group. Importantly, the mortality rate was relatively high in the simple complexity group who had exercise data. That group included aortic valve disease and aortic coarctation which are notable for the range of severity. By defining the baseline exercise group as >70% predicted workload, they do not examine those with the highest exercise capacity. It would be useful to understand if there was a group with negligible mortality. Many (maybe most) of the deaths in this cohort came in older patients with a mean age of 52 years. These data, most notable in figure 2, also remind the reader that for virtually all forms of 2 ventricle heart disease, there will be patients with supernormal and near normal exercise capacity.

The academic exercise community has some bias toward using cardiopulmonary exercise tests which provide elegant expired gas analyses and maximal and submaximal measures of oxygen capacity. Peak workload, as used here, is strongly correlated with those more elegant values. The technique has the advantage of requiring less expensive equipment and less training for the staff. The authors conclude that more adult CHD patients should get regular exercise testing. That conclusion is clearly valid. The authors also suggest that those evaluations are relatively simple, and they would benefit from looking at both the simplest and the more complex patients. That practice would help confirm or challenge our assessment of patient's physiology.

These data, of course, do not allow us to understand how much we are able to adjust those mortality curves, but by adding this information to our overall risk stratification assessment, we can advise our patients on the potential benefits of the intervention. I would hope, that by systematically measuring exercise capacity, we encourage the 71% of patients with <3 hours of activity/week, to increase their daily activity and achieve some of the benefits of exercise as shown in patients with other forms of heart disease.[4]

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