JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY
VOL. 65, NO. 18, 2015

ISSN 0735-1097/$36.00
PUBLISHED BY ELSEVIER INC.
http://dx.doi.org/10.1016/j.jacc.2015.02.054

EDITORIAL COMMENT

# Closure Is Not Correction

## Late Outcomes of Ventricular Septal Defect Surgery*

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Since the publication by Lillehei et al. of the seminal report of successful ventricular septal defect closure 60 years ago (1), dramatic improvements in surgical mortality and short- to medium-term outcomes have occurred (2–4). As a result of these improvements, mortality in infants and children with a ventricular septal defect has fallen 60% to 80% since 1979, and the prevalence of adults with the diagnosis of a ventricular septal defect is increasing (5,6). With a greater number of adults surviving decades after successful congenital heart surgery, the urgency for better characterization of the congenital heart disease–related morbidities and mortality that occur late after surgical intervention is growing. The report by Menting et al. (7) in this issue of the *Journal* adds important information to the limited data available regarding outcomes in late survivors of ventricular septal defect closure (8–11).

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The existing reports of outcomes in long-term survivors of ventricular septal defect closure span several eras of open heart surgery. The results of 30 and 53 years of follow-up of Lillehei's index cohort of patients who underwent closure between 1954 and 1955 using extracorporeal cross-circulation were published in 1986 and 2009, respectively (8,9). Because of the limitations in the size of the children for whom cross-circulation could be used effectively as a means of extracorporeal support, nearly one-half of the original cohort were operated on at age 1 year or less. Mortality in survivors of the early post-operative period was 10% over a 30-year period, with only 1 additional death occurring between 30 and 53 years after closure. In 1988, the Second Natural History Study reported on the outcomes of children with a ventricular septal defect operated on between 1958 and 1969 (11). Despite a decade of improvements in surgical techniques and myocardial preservation, 25-year mortality in survivors of the early post-operative period was 11%, similar to the cohort reported by Lillehei et al. In the current report, closure was performed even more recently (1968 to 1980); however, the mortality among survivors of the early post-operative period was surprisingly similar to the earlier reports (13% at 20 years) (7). Improvements in peri-operative surgical mortality do not appear to have translated into improvements in late mortality.

The current report highlights several areas that the cardiovascular specialist caring for adult survivors of ventricular septal defect closure must monitor carefully. Most important, arrhythmias and heart failure have a significant negative impact on late outcomes in patients who have undergone ventricular septal defect closure. Sudden death is an important mode of death in this population, and the identification and validation of a risk profile for tachyarrhythmias has the potential to identify patients who would benefit from primary or secondary prophylaxis with an internal cardioverter-defibrillator or medical therapy.

The progressive deterioration of biventricular cardiac function that was reported in the current cohort raises concerns that a significant proportion of these patients are at risk to develop heart failure in the decades to come. The ability of neurohormonal blockade to reverse the ventricular remodeling and prevent clinical heart failure is unknown in this population. There may be underlying differences in the myocardium of the patient who has undergone

*Editorials published in the *Journal of the American College of Cardiology* reflect the views of the authors and do not necessarily represent the views of *JACC* or the American College of Cardiology.

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ventricular septal defect closure that are not comparable to that of an adult heart with an ischemic or primary cardiomyopathy. The differences may affect the response to angiotensin-converting enzyme inhibition or beta-blockade. Use of beta-blockade in a patient with arrhythmias may also present challenges that require careful balancing of the risks and benefits before initiation and during therapy.

Once the patient was beyond the early postoperative period, late cardiac interventions in the current cohort were skewed toward treatment of left-sided heart lesions. Aortic insufficiency detected by echocardiography also appears to be progressive. It is difficult to ascertain from the data provided how many of the patients who developed left-sided lesions had left-sided disease apparent at the time of diagnosis and surgery. Nonetheless, the need for patients to have regular follow-up to monitor for associated lesions is clear.

The quality-of-life assessments and objective exercise testing results confirm the impressions of patients and cardiologists alike. Reported quality of life among the survivors in the current cohort was excellent and superior to the general population. Exercise performance was somewhat lower than expected for the general population but not enough to impact daily living. The observed deterioration in exercise capacity over time raises the possibility that exercise training and increased emphasis on fitness may be able to arrest or reverse this decline.

The current European Society of Cardiology recommendation for routine follow-up of a patient with no significant complications after ventricular septal defect closure is every 5 years (12). The American College of Cardiology/American Heart Association guidelines do not recommend follow-up at an adult congenital heart disease center for the patient with no residual ventricular septal defect, pulmonary hypertension, or associated lesions (13). This report demonstrates ongoing morbidity and mortality in adults who have undergone ventricular septal defect closure. The overall population of survivors is still so small that one would predict the exposure of a general cardiologist to a patient who has undergone a ventricular septal defect closure would be limited to a few patients. Until better risk-stratifying factors are identified, a strong argument could be made that even relatively straightforward patients such as those who have undergone ventricular septal defect closure should be followed up at an adult congenital heart disease center to ensure comprehensive evaluation and treatment as they age.

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**KEY WORDS** ventricular septal defect