SURGERY FOR CONGENITAL HEART DISEASE

EDITORIAL: INTRA-ATRIAL REPAIR OF TRANSPOSITION—LATE RESULTS AND MANAGEMENT PROBLEMS

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It is now 40 years since Senning¹ described an intra-atrial repair for babies with transposition of the great arteries. His operation, together with that subsequently performed by Mustard, transformed the prognosis of this previously fatal condition. Despite low operative mortality, however, these operations have been largely abandoned in favor of the arterial switch, because of concerns regarding long-term complications. Baffle obstruction, arrhythmia, and systemic ventricular failure have all been reported and suggested as important causes of late morbidity and mortality, particularly sudden death.^{2,3} The Senning operation was revived in the late 1970s and has been associated with less pulmonary and systemic venous pathway obstruction than the Mustard procedure. Nevertheless, the development of arrhythmia and progressive decline in the function of the systemic right ventricle have been considered inevitable by many groups. Risk stratification based on understanding of the etiology and functional significance of these late problems and the development of appropriate medical and surgical treatment strategies are challenges for those involved in the management of the large number of patients who have reached adulthood after having undergone the Mustard or Senning procedure.

In this issue of the Journal, Kirjavainen and colleagues⁴ describe a retrospective review of 100 patients who had undergone a Senning operation for both simple and complex transposition of the great arteries at a single institution with a mean follow-up of greater than

12 years. Their findings confirm many of those of previous reports but raise a number of concerns and management issues. Outlook was significantly worse for the patients with complex compared with simple transposition, emphasizing the importance of separating the two groups when studying long-term results. In both groups, there was a decreasing incidence of stable sinus rhythm with time. Although the rate of development of bradycardia, sinus node dysfunction, junctional rhythm, and other supraventricular rhythms varies according to the nature and intensity of testing, follow-up studies have been consistent in showing that most patients remain free of arrhythmic symptoms, that loss of sinus rhythm does not predict late sudden death, and that insertion of a pacemaker does not prevent sudden death.⁵ Kirjavainen and colleagues hypothesize that physiologic pacing may retard the development of ventricular dilatation and preserve systemic ventricular function, but this remains unproven.

The fate of the right ventricle in the systemic circulation remains uncertain, and the current report emphasizes the difficulties in studying this key aspect of cardiac function after intra-atrial repair. Qualitative assessment of systolic function by echocardiography is not optimal, and an appropriate "control group" for comparison and definition of "dysfunction" is not available. Like most previous reports, Kirjavainen and colleagues report a higher incidence of right ventricular problems at late follow-up than at earlier assessment. As serial evaluation was not undertaken, however, the inevitability of decline with time remains unproven. Indeed, Reich and colleagues,⁶ using radionuclide testing, have shown no deterioration in indices of systolic performance over a median interval of 8.8 years. Their data suggest that ventricular filling, rather than myocardial contractility, may be the limiting factor for cardiac output, and that this may deteriorate with time and growth. Kirjavainen and colleagues report only a small number of patients with tachycardias such as atrial flutter, which have been shown to be associated in previous

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studies with an increased risk of sudden death and hemodynamic decline.⁵ It is easy to understand how a tachyarrhythmia might be poorly tolerated in a circulation with compromised or limited ventricular filling. Patients who have palpitation or documented tachyarrhythmia should therefore have both careful electrophysiologic and hemodynamic evaluation. Further study is clearly warranted, and there are treatment implications. For example, the increasing use of angiotensin-converting enzyme inhibitors, as advocated in the current study for treatment or prophylaxis, is based on extrapolation from left ventricular function responses in adults with normally connected hearts.⁷ This approach may not be beneficial after Mustard or Senning operations and could be detrimental in some circumstances.

The management of those patients who have symptomatic severe right ventricular failure late after intra-atrial repair remains difficult. Late conversion to an arterial switch has been undertaken, but the mortality has been high.⁸ The good symptomatic status in the current and previous reports at medium-term follow-up makes this approach difficult to justify in well patients, particularly as the likelihood of developing clinical right ventricular problems cannot be easily predicted. It is also important to realize that the successfully switched circulation in such patients is not normal, with potential problems from ventricular remodeling, valve function, and cardiac rhythm. Heart transplantation has been performed and remains an alternative option in the small proportion of severely limited patients.

Current knowledge of the determinants of late morbidity and mortality after Mustard and Senning opera-

tions remains limited. Long-term prospective follow-up with protocols to assess the functional consequences of electrical and mechanical complications and their interaction should improve risk stratification, patient management, and counseling. The study by Kirjavainen and colleagues emphasizes the need for such meticulous long-term follow-up of patients after intra-atrial repair.

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