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EDITORIAL COMMENT

The Functionally Univentricular Heart

Which Is Better—Right or Left Ventricle?*

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The question regarding long-term outcome in patients with a functionally univentricular heart undergoing a Fontan procedure based on whether there is right or left ventricular morphological dominance is one that has been investigated for many years. The paper in this issue of the *Journal* by d'Udekem et al. (1) from the Royal Children's Hospital in Melbourne, Australia, found that in their nearly 30-year series reviewing 499 patients, right ventricular dominance was the single most important risk factor for death. Their other conclusion was that this risk factor seemed to be important only before the bidirectional superior cavopulmonary anastomosis (BSCPA). The interesting speculation is that earlier BSCPA may potentially modify the risk factor of right ventricular morphology. Both of these conclusions are important for clinicians to consider when guiding patients along a Fontan pathway, especially when considering timing of BSCPA and cardiac transplantation as an alternative.

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Their review (1) is a definite contribution to our understanding of the long-term outcome of patients undergoing the Fontan procedure for functionally univentricular heart. Certainly their conclusion that right ventricular dominance is a risk factor for death would be supported by an examination of patients undergoing the Fontan operation in the late 1970s and 1980s. In the interesting series of patients who have had the Fontan conversion at our hospital (N = 135), we have had the opportunity to see patients from these early years who had an atriopulmonary Fontan (2). Over 70% of patients who were able to survive long enough to have a Fontan conversion had a diagnosis of either double-inlet left ventricle or tricuspid atresia. In either of these diagnoses, the left ventricle was the predominant morphological ventricle. We have had very few patients in that historical series with right ventricular morphology who have survived an atriopulmonary Fontan and presented for Fontan conversion.

Some recent data from centers that have reviewed their results after the Fontan procedure for patients with hypoplastic left heart syndrome (right-dominant ventricular morphology) have clouded this question. In particular, 2 recent reports seem to contradict the conclusions of d'Udekem et al. (1). The 15-year single-institution experience from the group at the University of Michigan reported follow-up on 636 primary Fontan procedures (3). One of the conclusions was that the Fontan procedure can be performed with low risk regardless of ventricular anatomy. The mean follow-up in that series, however, was only 15 months (range 0 to 173 months). The group from Milwaukee reported 256 patients undergoing a Fontan procedure and also concluded that "ventricular morphology did not predict outcome" (4). In that series, the mean event-free survival at 10 years was 75 ± 7% in the left ventricular group and 67 ± 9% in the right ventricular group. The results of these 2 groups were not statistically significantly different, but the results may be evidence of a trend agreeing with the conclusion of d'Udekem et al. (1) that right ventricular dominance is a predictor of earlier mortality.

A confounding issue with all of these studies is the length of follow-up. However, if one turns to other congenital cardiac lesions that have right ventricular dominance, the evidence for eventual (premature) failure of the right ventricle becomes more apparent. If one looks at patients with congenitally corrected transposition of the great arteries, it seems that in these patients there is an inexorable decline in right ventricular function over time. In the landmark review by Presbitero et al. (5), congestive heart failure developed in 25% of patients in the fifth decade of life and in >60% of patients in the sixth decade of life. Significant arrhythmias developed in 50% of patients by the fifth decade of life and in 80% of patients by the sixth decade of life. In the Toronto review, the mean age at death in patients with congenitally corrected transposition and no other cardiac defects was 38.5 years (6). These patients died of right ventricular failure and sudden death. The other group of patients who have had the right ventricle acting as the systemic ventricle are patients who have undergone the atrial baffle procedure for transposition of the great arteries. In the review by Gelatt et al. (7), arrhythmias developed in 75% of patients at 20 years after the Mustard operation. The estimate of late survival at hospital discharge after the Mustard procedure was 70% at 25 years.

I agree with the conclusion of d'Udekem et al. (1) that right ventricular dominance is indeed an important risk factor for death after the Fontan procedure. I believe this finding is supported not only by their excellent analysis but

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by historical data at our institution from the group of patients who had an atriopulmonary Fontan procedure, the deterioration of the right ventricle in patients with congenitally corrected transposition of the great arteries, and the decrease in ventricular function in patients undergoing an atrial baffle operation for transposition of the great arteries. Additional support for this conclusion is provided by the groups from the Mayo Clinic (8) and Birmingham, United Kingdom (9). Julsrud et al. (8), in reviewing 500 Fontan patients, concluded that left ventricular morphology was associated with a better early survival than right ventricular morphology. McGuirk et al. (9) found that a morphological right ventricle was a risk factor for prolonged in-hospital stay, which influences long-term survival.

A secondary issue raised by d'Udekem et al. (1) is the observation that right ventricular dominance as a risk factor for death seemed to be important only before the BSCPA. A possible implication here is that performing the BSCPA sooner may in fact decrease the overall risk of mortality. I am not sure this is completely supported by the evidence. The loss of patients in the early time period may actually be a force of natural selection. If the child can survive until the time of the BSCPA, that outcome may be a result of natural selection of the patients who are more fit. It will take longer follow-up to show that early BSCPA is, in fact, a modifier of outcomes in patients with right ventricular dominance.

I congratulate the authors on an extremely thorough review of a large number of consecutive patients from a single institution. This research will help us with our clinical decision making in borderline patients when we are trying to decide between continuation along the Fontan pathway versus cardiac transplantation. All other factors being equal, patients with right ventricular dominance and other risk factors for poor outcome after the Fontan procedure may be ones that should be directed toward cardiac transplantation sooner rather than later.

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Key Words: bidirectional Glenn ■ Fontan ■ functionally univentricular heart ■ left ventricle ■ right ventricle ■ single ventricle.