Unsatisfactory Early and Late Outcomes After Fontan Surgery Delayed to Adolescence and Adulthood

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The ideal age to perform the Fontan procedure is still unknown. The aim of this study is to determine outcomes after Fontan surgery delayed to adolescence and adulthood in Australia and New Zealand. Patients who had undergone a Fontan procedure at 15 years of age or older were identified in the 1133 patients registered in the Australia and New Zealand Fontan Registry until December 2012. A total of 45 patients underwent the following Fontan procedure at a median age of 18.3 years (16-21 years): 24 atriopulmonary connections, 10 lateral tunnel, and 11 extracardiac conduits. Hospital mortality was 13% (6 of 45). After a mean follow-up of 15.5 ± 9 years, there were 8 late deaths. Survival rates after 10, 20, and 25 years were 79% (95% CI: 64-89), 70% (95% CI: 51-83), and 70% (95% CI: 51-83), respectively. Freedom from Fontan failure (death, heart transplantation, Fontan takedown, protein-losing enteropathy, and poor functional status) after 10 and 20 years was 63% (95% CI: 47-76) and 35% (95% CI: 19-52), respectively. Patients with a single left ventricle had a lower risk of failure (hazard ratio = 0.25, 95% CI: 0.10-0.59; $P = 0.002$). Arrhythmias developed in 29 patients after a median of 0.1 years (0-9.3 years) and 10 required a permanent pacemaker. Freedom from all adverse events at 10 years was 30% (95% CI: 16-45). Outcomes of the Fontan procedure in adolescents and adults are poor, with disproportionately high hospital mortality and late adverse events. The Fontan procedure should not be delayed to adolescence and adulthood and should be performed electively in childhood.

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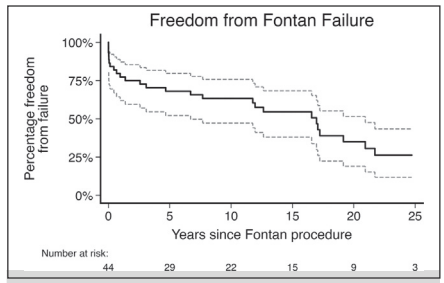

High rate of failure when Fontan surgey is delayed after 15 years of age.

Central Message

Mortality and failure are high when the Fontan procedure is performed after the age of 15 years. Fontan surgery should not be delayed after adolescence.

Perspective

The Fontan circulation is still doomed to fail, and some believe that waiting as late as possible to perform the procedure is beneficial. In a retrospective review of 45 patients operated after the age of 15 years, we demonstrated that hospital mortality and rate of failure were much higher than expected. We therefore believe that Fontan surgery should not be delayed to adolescence.

See Editorial Commentary pages 175–176.

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INTRODUCTION

It is more than 40 years since the first description of the Fontan procedure, and the ideal age to perform the procedure is still unknown.[1-7] Although most the centers worldwide are performing the procedure in infancy between 2 and 4 years of age, some teams advocate to wait to a later age.[5,6] The rationale to postpone the procedure is based on the belief that the Fontan circulation provides adequate palliation for a limited time and that postponing this surgery may improve longevity.

In recent years, the teams who have reported their experience with adults and adolescents undergoing Fontan surgery have demonstrated their hospital mortality to be between 4% and 13%, higher than the 1% usually reported recently for this surgery when performed at a younger age.[1,3,8-15] However, more importantly, in limited follow-up extending to 15 years, outcomes seem to be equivalent to those achieved by younger patients.[1-3,5,8-10]

The Australia and New Zealand Fontan Registry has been created to prospectively gather follow-up information of all patients in the region who have undergone a Fontan procedure.[16] We decided to review our regional experience with the Fontan procedure performed in adolescents and adults to investigate the late outcomes of this population.

METHODS

The Australia and New Zealand Fontan Registry collects data on patients who underwent their Fontan procedure locally, as well as patients who had their operation overseas but are now followed up in either country. The organizational structure of the Registry has been described previously.[16] Ethics approval was granted nationally in both Australia and New Zealand by the institutional review board of all participating institutions. Written or verbal consent to participate in the Registry was obtained prospectively following its establishment in 2008 and retrospectively for patients who underwent their operation before 2008, including those who died or required heart transplantation. A total of 13 centers (7 pediatric and 6 adult) overseeing the care of patients with Fontan circulations in Australia and New Zealand participated in the study. Follow-up data were extracted from hospital inpatient records, outpatient clinic attendances, and private cardiology clinical summaries. For those patients who were dead or who declined to participate in the Registry, ethics approval was granted for the retrospective audit of their health records. There is central collection of all death information in New Zealand, and an audit of the National death Index of Australia was performed in 2013, ascertaining that patients who did not present recently to follow-up are not dead.

A total of 1133 patients who underwent the Fontan procedure were registered in the Australia and New Zealand Fontan Registry as of December 2012. Patients were excluded from the study if they were referred to the Registry from overseas or underwent their Fontan operation before 15 years of age. The 45 patients within the Registry who underwent their Fontan operation at 15 years of age or older constitute the cohort of this study. Follow-up data were 87% complete in this cohort. Of the 45 patients included, 24 had an atriopulmonary connection (AP; 1975-1996), 10 had a lateral tunnel (LT; 1990-2000), and 11 had an extracardiac conduit (ECC; 1999-2006).

Definitions

Early mortality was defined as death within 30 days of the Fontan procedure or before hospital discharge. Late mortality was defined as death after 30 days and hospital discharge. Prolonged effusions were defined as pleural effusions lasting for more than 30 days or requiring pleurodesis. Fontan failure was defined as late death, heart transplantation, Fontan takedown, Fontan conversion from an AP or LT to an ECC, protein-losing enteropathy, plastic bronchitis, moderate to severe left ventricular (LV) dysfunction, or New York Heart Association (NYHA) class III or IV. Thromboembolic events were defined as thrombus within the Fontan circulation, pulmonary embolism, transient ischemic attack or stroke, or thrombus in the venous circulation. An arrhythmia was defined as the occurrence of supraventricular tachycardia, complete heart block, ventricular tachycardia, or ventricular fibrillation. An adverse outcome was defined as the presence of Fontan failure, a thromboembolic event, or arrhythmia or the insertion of a permanent pacemaker.

Statistical Analysis

The end points examined for the 45 patients who underwent the Fontan procedure were (1) mortality, (2) first thromboembolic event, (3) first arrhythmia, (4) Fontan failure, (5) time to occurrence of moderate to severe LV dysfunction, and (6) occurrence of a composite adverse event outcome. In addition to the characteristics described in the Table, surgery in the era before 1991 was analyzed as covariate, as was year of Fontan procedure completion. Analysis of type of Fontan was limited to contemporary techniques (LT and ECC) vs classical AP Fontan. Overall survival and freedom from each of the nonmortality end points were examined using Kaplan-Meier