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ORIGINAL ARTICLE

Extracardiac Conduit Fontan – Outcome Data in Early Adulthood

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Q4 **Background** To describe the survival and health outcome status of young adults with an extracardiac Fontan procedure performed either as a primary or conversion (secondary) Fontan surgery.

Q5 **Methods** The database of the Adult Congenital Heart disease service at the Royal Melbourne Hospital was interrogated to identify all adults who had undergone a primary extracardiac conduit Fontan (n = 29) or a Fontan conversion with this procedure (n = 8). We then determined vital status, age, original anatomy and functional status in early adulthood in both groups.

Results Adults with an ECC Fontan procedure report reasonable NYHA functional class (84% NYHA I or II) though, objectively, exercise testing demonstrates a reduced exercise capacity, and desaturation on exertion is frequent. The majority (86%) have completed secondary education. Most (78%) are managed on warfarin and there is a preponderance of ACE inhibition use (62%). Atrial arrhythmias have been documented in 5 of the 29 primary ECC groups (17%); in 3 patients this preceded primary ECC and 2 patients developed post primary ECC (between 6 and 14 years postoperatively). At a lesser time of follow-up [median 4.5 years (IQR 3.3–6)], conversion to an ECC as a secondary Fontan procedure has successfully treated atrial arrhythmias in the 7 (of 8) patients where this was the surgical indication for conversion.

Conclusions Though long-term data will require decades to establish, in young adulthood the functional outcomes of a primary ECC Fontan operation are encouraging. Secondary ECC conversion successfully mitigates atrial arrhythmias in the short to medium term.

Keywords Congenital heart defect • Fontan procedure • Young adult • Survival • Exercise test • Echocardiography

Introduction

Q6 A Fontan circulation is the final destination surgery for children with cardiac anatomy not suitable for a two-ventricle repair. The most recent version of the surgically created Fontan circulation is the extra cardiac conduit (ECC) [1], with surgical innovation driven by recognition of the propensity for clot formation, atrial arrhythmias and energy loss within the original atriopulmonary Fontan connection.

The time required to ascertain long-term sequelae of refinements in surgical approach, makes determination of outcomes an ongoing priority in adults with congenital heart disease. Additionally, in patients with earlier Fontan surgery, conversion to an ECC may occur in adulthood, with the commonest indication being atrial arrhythmias. How, and whether, this Fontan conversion will impact long-term outcomes remains unclear. Recent publication of local data from the Australia and New Zealand Fontan Registry (ANZ

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Registry) shows significant early mortality with this approach, though postoperative outcomes are better in those who undergo conversion earlier in life [2].

Available literature suggests that individuals with an ECC Fontan may have less arrhythmia over the short term [3,4]. Medium-term follow-up in more recent studies has not determined any difference in arrhythmia incidence [5,6]. ANZ Registry data is encouraging, but again limited in only showing data to approximately 15 years post Fontan surgery in the ECC cohort [7]. Questions remain about the long-term benefit of this approach with respect to arrhythmia incidence and particularly the challenges of subsequent treatment, if required, with this circuit [8,9]. Locally, surgical adoption of the ECC Fontan connection commenced in 1998 and became the exclusive Fontan procedure in 2006 [10].

To date, within the population of Fontan circulation patients, worse outcomes have been demonstrated for those with hypoplastic left hearts (HLHS) [7]. There is little long-term data on this particular subgroup of Fontan patients. Proportionally, they are over-represented within the ECC cohort as compared with other Fontan operations as they have become eligible for a Fontan procedure more recently than individuals with other anatomical diagnoses.

In view of the questions that remain about longer-term outcomes of patients with ECC Fontan circulation, those with an anatomical diagnosis of HLHS and adults who have undergone ECC conversion, we sought to ascertain the outcomes in our cohort of patients in early adulthood. Ethics approval was granted by the Human Research and Ethics Committee of the Royal Melbourne Hospital (QA2015015).

Aims

To identify all patients of the Royal Melbourne Hospital Adult Congenital Heart Disease service who have undergone either a primary or secondary ECC Fontan operation and determine their outcomes in adulthood, in particular: clinical functional status, systemic ventricular function, anticoagulation and medication use, occurrence of complications (including arrhythmias), and self-reported functional capacity and educational level attained. Patients had to have been referred by the end of 2014 for inclusion.

To describe and compare outcomes between those with a primary and those with a secondary ECC; and to describe outcomes between those with a HLHS versus other anatomical diagnoses.

Methodology

The database was searched for all patients entered as having undergone a Fontan operation. Patients with an extracardiac Fontan connection were identified from surgical details and denoted to be a primary ECC Fontan patient where this was the first Fontan surgery or a secondary ECC patient where conversion had occurred from either an atriopulmonary (AP) or lateral tunnel (LT) Fontan connection.

Data is presented as median (interquartile range).

Variables described are: age of patient, original anatomical diagnosis, years post Fontan surgery, rhythm documented on most recent ECG, echocardiogram, oxygen saturation (pre and post exercise test), functional testing (exercise stress test or 6-minute-walk-test (6MWT)), self-reported functional status, systemic ventricular function, medication and anticoagulation use and complications (thromboembolic or arrhythmic).

Results

Cohort Characteristics

We identified a total of 37 ECC Fontan patients, of whom 29 had undergone a primary and 8 a secondary ECC Fontan operation. Their median age at the last clinic visit was 22 years (20–26). As expected, the secondary ECC Fontan patients were older, aged 27 (24.3–32) at their last clinic review. Of the 37 patients, 33 were in regular care at the time of data ascertainment. Previously, 1 patient had undergone cardiac transplantation for refractory heart failure and 1 patient had committed suicide. Two patients had been lost to follow-up, defined as last visit >2 years previously. Of these, 1 patient has again agreed to attend clinic (there have been multiple telephone contacts over the years with repeated failures to attend).

One patient unfortunately died in hospital following a respiratory arrest complicating a cerebral infarct as this manuscript was being prepared. That patient had been anticoagulated with warfarin, but had recently been subtherapeutic in the setting of a procedure and antibiotic use with cautious warfarin prescribing (INR 1.6 at presentation).

The cohort is a median of 13 (6–14) years post Fontan operation, with the primary ECC having a median of 13 (11–15) years of follow-up and the secondary ECC having a median 4.5 (3.3–6) years of follow-up. ECG at last follow-up recorded: sinus rhythm (n = 29), sinus bradycardia (n = 2) or a paced rhythm (n = 5) (missing: patient post heart transplant whose follow-up has been at another institution).

Anatomical Diagnosis and Operative Details

The age at the time of ECC Fontan operation differed significantly as expected. The primary ECC Fontan operation patients had surgery at 8.3 years of age (5.2–14.5) and the secondary ECC Fontan operation patients at 20.5 years of age (18.2–27.4) (Table 1).

Self-Reported Physical/Functional and Educational Status

Self-reported functional status, reflected by NYHA classification based on self-reported activity is reasonable. The majority of the cohort (31, 84%) is NYHA class I (n = 17) or NYHA class II (n = 14).

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