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ORIGINAL ARTICLE

Long-term Follow-up of Adults Following the Atrial Switch Operation for Transposition of the Great Arteries – A Contemporary Cohort

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Background The atrial switch operation for transposition of the Great Arteries (TGA) (Mustard or Senning Procedure) provides excellent short-term survival. Significant long-term concerns exist for these patients, however, including the ability of the right ventricle to maintain systemic perfusion and the risk of arrhythmia. We seek to describe long-term mortality and morbidity of this group of adult patients.

Methods Consecutive patients who had undergone an atrial switch procedure, who were aged over 16 years and who were followed up at our tertiary level adult congenital heart disease (ACHD) service in Sydney, Australia since 2000 were included. We documented mortality using a National Death Index and analysed the prospectively defined composite endpoint of ''Serious Adverse Events'' including death, heart failure hospitalisation and/or documented ventricular arrhythmia.

Results There were 83 patients included; mean age at most recent follow-up was 35 ± 5 years. Overall survival was 82% at 35 years and 22% of patients experienced a serious adverse event. Atrial and ventricular arrhythmias occurred in 45% and 7% of patients respectively. Eighteen (22%) patients required a pacemaker and six (7%) required implantable cardio-defibrillator (ICD) implantation. Significant right ventricular dysfunction was present in 26% of patients on their most recent visit and this, or requirement for permanent pacing, was associated with an increased risk of serious adverse events (OR 10.22, $p < 0.001$), (OR 4.998, $p = 0.04$) respectively.

Conclusions Significant mortality and morbidity accrues by mid-adult life after an atrial procedure for TGA. Right ventricular dysfunction and permanent pacing are associated with serious adverse events.

Keywords Transposition of the great arteries • Congenital heart disease • Atrial switch

Introduction

Transposition of the Great Arteries (TGA) is a congenital heart defect where the pulmonary and aortic trunks are supplied by the incorrect ventricles. The atrial switch by the ''Senning'' [1] or ''Mustard'' [2] procedure became the standard ''complete'' palliative operation for patients with dextro-Transposition (d-TGA) from the mid 1960s to the mid

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1980s, when the use of the arterial switch procedure became commonplace. These operations revolutionised a condition that otherwise had a 90% mortality rate in the first year of life [3] and provided very good short-term outcomes, into late childhood and early adulthood [4]. The development of significant late complications is common, however, including systemic ventricular dysfunction, need for re-operation, arrhythmias, heart failure and sudden cardiac death [5–9]. Patients with previous Mustard or Senning procedures are now approaching middle age and longer term follow-up is thus now possible. Understanding the course of a cohort of adult patients will enable more appropriate risk stratification and prognostication for patients, families and physicians. Data on patients heading to their third and fourth decade of life is limited. We therefore aimed to assess outcomes, in our cohort of adult TGA patients.

Methods

Study Patients and Definitions

A single-centre retrospective study of prospectively collected data was performed at a tertiary level adult congenital heart disease (ACHD) referral centre in Sydney, Australia. This is the largest ACHD referral centre statewide, with a 'catchment area' of six to seven million people.

Consecutive patients who had undergone a Mustard or Senning repair for "simple" d-TGA were included if they were ≥16 years and had been seen at least once at our centre since January 2000. "Simple" TGA was defined as d-TGA without other cardiac abnormality including (other than small) ventricular septal defect (VSD), pulmonary stenosis, double outlet or single inlet ventricles.

Operation reports and clinical notes were reviewed for additional cardiac abnormalities, interventions or operations prior and subsequent to the atrial switch operation. Postoperative course including arrhythmias, devices, functional status, need for heart transplant and complications occurring before admission to the ACHD clinic was recorded on initial clinic visit and then prospectively acquired from entry into our service.

Arrhythmias were included if documented via electronic medical record, electrocardiogram, implantable device checks or if the patient had symptoms suggestive of an arrhythmia which was inducible on electrophysiology study and which was clinically relevant.

Echocardiography for initial and most recent clinic visit were completed and reviewed. Systemic right ventricular function was evaluated by qualitative assessment of clinic echocardiograms. Ventricular function and tricuspid regurgitation were graded by experienced ACHD physicians as either normal, mild, moderate or severe dysfunction. Pulmonary hypertension was assessed and classified as per current guidelines [10].

Systemic venous baffle obstruction was diagnosed as low flow velocity (<0.5 m/s) in either the inferior vena cava or superior venae cava flows with flow >1.0 ms in the baffle. Pulmonary venous baffle obstruction was diagnosed if the peak velocity at the narrowest point of the baffle was greater than 1.5 m/s.

Survival and Adverse Event Outcomes

Survival data were obtained for every patient from the National Death Index (NDI), last clinic visit and cross-referenced with clinic visit records. Cause of death information was reviewed using medical record information, death certificates and autopsy information where available. A composite endpoint of "Serious Adverse Events" was prospectively defined as all-cause mortality, documented ventricular arrhythmia, heart failure requiring hospitalisation and/or the need for defibrillator implantation. The development of severe pulmonary hypertension and the need for heart transplantation was documented.

Ethics approval was provided by Sydney Local Health District (SLHD) Research Ethics and Governance Office.

Statistical Analysis

Statistical analysis was completed using Statistical Package for Social Services version 22.0 (SPSS, Inc. Chicago IL). Continuous variables are presented as mean ± standard deviation or median with range. Student t-test was used for comparison of continuous variables. Categorical variables are presented as frequencies and percentages. Comparison of categorical variable groups was performed using chi-squared test or Fisher

Table 1 Baseline Characteristics.

Variable	All Patients (n = 83)	Patients with Serious Adverse Event Outcome (n = 18)	No Serious Adverse Event Outcome (n = 65)	p value
Mean age at initial repair	1.4 ± 2.7	1.6 ± 2.1	1.3 ± 2.9	0.73
Mean age at last follow-up	35.1 ± 5	36.2 ± 6.6	33.1 ± 5.4	0.05
Male gender, n (%)	61 (74)	15 (83)	41 (63)	0.10
Time from first ACHD visit to last follow-up	10.1 ± 6.1	10.6 ± 8	10 ± 5.5	0.70

Abbreviation: Adult Congenital Heart Disease.

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