Outcome of atrial repair procedures in patients with transposition of the great arteries followed up in a district general hospital

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Abstract

Patients with transposition of the great arteries (TGA) are now living longer due to improved medical and surgical care. Most of the current patients with TGA followed up at our district general hospital (DGH) grown-up congenital heart (GUCH) clinic have undergone a Mustard or a Senning atrial repair procedure between the early 1960s to mid 1980s. Complications found to be associated with the atrial repair procedure include arrhythmias, right ventricular impairment, tricuspid valve dysfunction, baffle-related problems and sudden death. This article reviews the outcome of patients with TGA in this DGH population and also addresses the issue of pregnancy and insurance.

Key words: outcome, atrial repair, transposition, great arteries, district general hospital.

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Introduction

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart condition presenting in the neonate. The overall annual incidence is 20–30 per 100,000 live births. It is characterised by the right ventricle functioning as the systemic ventricle with the tricuspid valve serving as the systemic atrioventricular valve. If left untreated, there is up to 90% mortality by the end of the first year of life.[1]

Between the early 1960s and the mid 1980s, the Mustard and Senning procedures were treatments of choice for patients with TGA. The Senning procedure, developed in 1958, directs the venous return to the contralateral atrioventricular valve and ventricle by means of an atrial baffle made of the patient's septal tissue. The Mustard technique, introduced later in 1964, excises the atrial septum and a pericardial or synthetic baffle is used to direct venous return.[2] These procedures were usually performed between one month and one year of age.

Due to complications associated with atrial repair procedures, including right ventricular failure, arrhythmias, tricuspid valve dysfunction, baffle-related problems and sudden death,[3] these procedures have been replaced by arterial switch operations (Jatene) since the mid 1980s. Most of the adult patients with TGA currently followed up in this GUCH clinic, however, have undergone the atrial repair procedure.

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Methods

Details of patients followed up at the GUCH clinic in this district general hospital are held in a database. Using this database, we sourced patients with TGA who had undergone an atrial repair procedure. Their medical notes were retrospectively reviewed for late complications related to the surgery, looking specifically for right ventricular impairment, arrhythmias, tricuspid valve dysfunction, baffle-related problems and sudden death. Simple TGA included patients without concomitant anomalies or a ventricular septal defect not requiring closure. Complex TGA included patients with other associated cardiac lesions including large ventricular septal defects, pulmonary stenosis and coarctation of the aorta. Systemic ventricular function was assessed routinely using two-dimensional (2D) echocardiography. Where magnetic resonance imaging (MRI) had also been performed, the ejection fraction MRI result was used instead of the 2D echocardiograph.

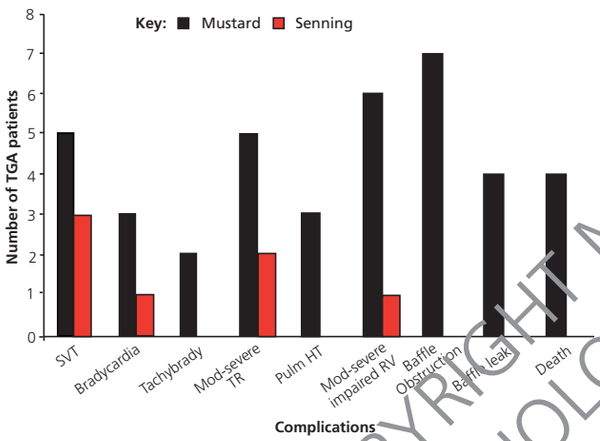
Results

A total of 30 patients with a diagnosis of TGA were followed up at the GUCH clinic: 22 of the 30 patients had previously undergone an atrial repair procedure. Two of the 22 patients were excluded as their care had been taken over by other hospitals. The other eight patients had either undergone an arterial switch, a Rastelli operation or had a congenitally corrected transposition. The male to female ratio was 2.3:1. The age of patients followed up (either until 2005 or until age of death) ranged from 16 to 46 years. Mean age of follow-up was 29 years. One of the 14 patients had undergone a redo Mustard performed three years later. None of the patients had undergone another type of atrial repair or arterial switch procedure performed after initial operation. Table 1 identifies the patient characteristics and figure 1 the major complications of this patient group.

Table 1. Characteristics of patients who underwent atrial repair for transposition of great arteries (TGA)

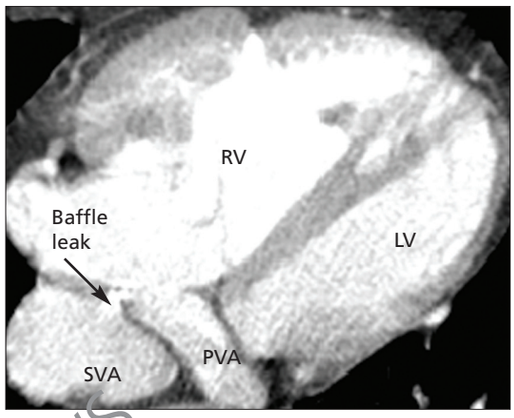
	Number of patients (%)	Mustard	Senning
Men	14 (70%)	9	5
Women	6 (30%)	5	1
Simple TGA	10 (50%)	6	4
Complex TGA	10 (50%)	8	2
Total	20	14	6

Figure 1. Comparison of complications amongst patients undergoing the Mustard or Senning procedures



Key: TGA = transposition of the great arteries; SVT = supraventricular tachycardia; mod = moderate; TR = tricuspid regurgitation; pulm = pulmonary; HT = hypertension; RV = right ventricle

Figure 2. CT scan of the heart showing a baffle leak



Key: RV = right ventricle; LV = left ventricle; SVA = systemic venous atrium; PVA = pulmonary venous atrium

Some 40% of patients with TGA had atrial flutter /fibrillation or ectopic atrial tachycardia. Half of our patients undergoing the Senning procedure had a supraventricular tachycardia (SVT) – one patient underwent ablation, one patient is awaiting ablation and one patient had a pacemaker inserted. Four patients had bradycardia or sinus pauses on Holter monitoring. Two of the patients required a pacemaker.

We found 35% of TGA patients, who had all undergone Mustard repair, had baffle obstruction. Three of the seven patients had dilatation with or without stenting of obstruction. Some 20% of TGA patients, all Mustard patients, had a baffle leak (see figure 2). One of the four leaks was closed.

Of the four deaths, one death was secondary to an arrhythmia, one died from an intervention-related cause and two died from secondary causes to pulmonary hypertension – one of which was secondary to pulmonary embolism on necropsy. All four deaths were in patients who had undergone Mustard repair and three were patients with complex TGA. There were no deaths in patients who had undergone the Senning procedure. Age on death ranged from 21 to 35 years. Two of the six female patients had successful pregnancies. Four of our patients had either learning or behaviour difficulties with or without epilepsy.

Discussion

Survival in patients with TGA is good with a 90% 10-year and an 80% 20-year survival rate but sudden cardiac death has been found to occur in 7–15% of patients with TGA.[4] The most common reason for death were sudden rhythm disturbances and systemic right ventricular dysfunction.[5] Hence, it is important to perform regular Holter monitoring and echo/MRI to review systemic ventricular function. In our patient group, 20% of the patients died during late follow-up. Three of the patients who died were known to have atrial flutter and one had sinus rhythm with short junctional rhythm on Holter monitoring. Kammeraad *et al*.[6] found the presence of symptoms of arrhythmia and heart failure and the presence of documented atrial flutter/fibrillation to be associated with a risk of sudden death. Sarkar *et al*.[7] reported that documented atrial flutter /fibrillation resulted in a 21-fold increase in the risk of late sudden death whilst Gelatt *et al*.[8] could not find a relationship between incidence of atrial flutter/fibrillation and sudden death. There is evidence in the paediatric population that the development of junctional rhythm may be a precursor of flutter or sudden cardiac death. Bradycardia has not been found to be a risk factor for mortality in previous studies. None of our Senning patients have died but their ages range from 16–25 years. A study by Moons *et al*.[9] found that Senning patients had a non-significant slightly better survival rate than Mustard patients.

At least one episode of arrhythmia was found in 70% of our patients with TGA. SVT was the most common, occurring in 50% of Senning patients and 36% of Mustard patients.

It is known that MRI is the ideal method to assess right ventricular systolic function but in view of the time it takes, its expense and its inability to be used in patients with pacemakers, it is not routinely performed on all our patients. A quarter of our patients with TGA had a normal systemic ventricular function. There was a lower proportion of our Senning patients with an impaired right ventricle and this may have been due to this group of patients being younger. An indicator of deterioration of right ventricular function is an increase in QRS duration.[10] Development of subpulmonary dilatation and failure should alert the physician to the possibility of pulmonary hypertension. Early aggressive therapy may delay transplantation, increase survival and improve quality of life.[11]

Moderate-to-severe tricuspid regurgitation (TR) occurred in 36% of Mustard and 33% of Senning patients. Mild-to-moderate TR is relatively common after atrial repair. As a result of the reversal of ventricular pressure, there is an alteration to the geometry of the ventricular septum that leads to the tricuspid valve developing a more rounded shape. There is then a tendency for TR due to displaced septal chordal attachments and the rounded tricuspid valve shape.

The finding that 20% of our TGA patients had a baffle leak is in keeping with other studies which have found this to be present in up to 25% of patients with TGA.[12] Learning difficulties with and without epilepsy were found in 20% of our TGA patients and 75% of them had undergone a Mustard procedure. This may reflect an extended period of hypoxaemia peri-operatively and during infancy.

Pregnancy

Pregnancy can cause a deterioration in New York Heart Association (NYHA) functional class, right ventricular dysfunction and atrioventricular regurgitation that is sometimes irreversible.[13] Other complications include clinically significant arrhythmias, especially SVT (associated with history of arrhythmias), miscarriage, premature delivery and an 11.8% fetal and neonatal mortality rate.[14] Pre-pregnancy exercise testing will allow assessment of functional capacity. The ability to complete nine minutes of a Bruce protocol suggests the likelihood of a good maternal outcome. Concomitant oxygen saturation with exercise should also be monitored since a resting oxygen saturation of less than 86% or a significant fall with exercise may compromise fetal well-being and lead to fetal loss.

Siu *et al*.[15] cared for 25 pregnancies in women with atrial repairs over a five-year reporting period, of whom six had complications which included heart failure, arrhythmias and death from post-partum heart failure in one patient (4% mortality). The paper also provided some guidelines of the likely complications during pregnancy in patients with congenital heart disease. This involved a rating system where one point was scored from each of four domains: i) prior cardiac event (heart failure, transient ischaemic attack, stroke before pregnancy) or arrhythmia; ii) baseline NYHA class>II or cyanosis; iii) left heart obstruction (mitral valve area<2 cm^2; aortic valve area<1.5 cm^2 or peak left ventricular outflow tract gradient > 30 mmHg by echo); iv) reduced systemic ventricular ejection < 40%. A score of one point indicated a 25% likelihood of maternal complications but a score of two points increased the risk massively to 75%. Nevertheless, pregnancy in patients with TGA is usually well tolerated with a systemic ejection fraction > 40%, NYHA I/II and unobstructed venous pathways.[16,17]

Key messages

- TGA (transposition of the great arteries) is the most common form of cyanotic congenital heart disease and therefore likely to be seen by adult cardiologists
- > 80% survive more than 20 years and into adulthood. On-going follow-up by cardiologists with training in adult congenital heart disease is important
- Palliation of simple TGA historically has been with the Mustard or Senning procedure ('atrial' repair)
- Late complications of atrial repair include rhythm disturbances which may indicate pathway (baffle) obstruction or systemic (right) ventricular failure
- There is unpredicted late sudden cardiac death in 7–15% but, generally, TGA is an intermediate insurance risk
- Carefully supervised pregnancy is possible; inheritance risk is very low in TGA
- Arterial switch is now the preferred intervention – late complications of which are now being seen

Insurance

Due to more effective medical and surgical care now being available, an increasing number of patients with TGA are surviving into adulthood and will, therefore, be seeking insurance cover. Health insurance usually does not cover costs associated with pre-existing conditions and is usually denied to patients requiring regular cardiac follow-up.[18] Adults with congenital heart disease are significantly more likely to have difficulty obtaining life insurance or mortgages. A questionnaire done by Crossland *et al*. found refusal rates to be independent of severity of congenital heart disease.[19] Vonder Muhll *et al*.[18] found that patients with TGA atrial repair are an intermediate prognostic group, indicating that they may be able to achieve insurance on an individual consideration, especially if they have no negative prognostic features which include early surgical era, ventricular dysfunction, arrhythmias and pulmonary hypertension. Some of their helpful suggestions include getting advice from a local adult congenital heart disease patient association (www.guch.org.uk) about insurers known to provide cover to GUCH patients, the importance of shopping around various insurers, getting a non-renewable term policy which

provides short-term coverage for a fixed period, and adoption of good health practices and avoidance of smoking.

Conclusions

Atrial repair procedures for patients with TGA have been found to be associated with complications including right ventricular failure, arrhythmias, tricuspid valve dysfunction, baffle-related problems and sudden death. We advise that patients should be screened for complications at yearly reviews by 12-lead electrocardiogram, regular Holter monitoring and echocardiography to look at baffle obstruction or leak and systemic right ventricular function. Identifying patients at risk of sudden cardiac death continues to be difficult. Only by systematically recording information may we learn about pointers to help prevent this in the future. Due to improved medical and surgical care, patients are living longer which requires us to address issues such as pregnancy and insurance.

Conflict of interest

LJF has received support attendance at meetings from Actelion. SR: none declared.

Editors' note

An editorial on 'Adult congenital heart disease: follow-up and the role of the district general hospital' by Drs DJ Gwilt and MA Gatzoulis can be found on pages 5–7 of this issue.

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