# Long-term results of anatomic correction for congenitally corrected transposition of the great arteries: A 19-year experience

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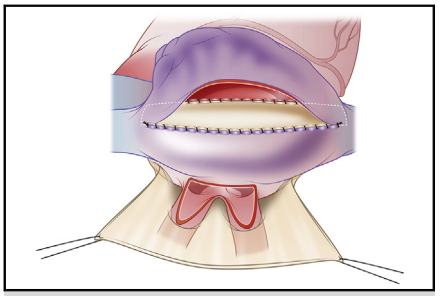
## ABSTRACT

**Objective:** The surgical indication, timing, strategy, and surgical technique for anatomic correction of congenitally corrected transposition of the great arteries are challenging. We evaluated the long-term results at The Royal Children's Hospital Melbourne.

**Methods:** Review of 32 successive anatomic corrections between 1996 and 2015.

**Results:** Twenty-one double-switch (66%), 6 Senning/Bex-Nikaidoh (19%), and 5 Senning/Rastelli (16%) procedures were performed (median age, 1.9 years). Median follow-up was 5.4 years with 4 deaths and 1 heart transplant. Cumulative incidence of late reoperation was 8%, 29%, and 59% at 1, 5, and 10 years, respectively. Twenty-six patients had full follow-up with native hearts. Nineteen had normal left ventricle (LV) function. Late LV dysfunction, mostly mild, was not related to needing a pacemaker ($P = .4$) or a pulmonary artery band (PAB) ($P = .08$). Previous PAB was linked to the need for aortic valve surgery or neoaortic regurgitation moderate or greater ($P = .03$). Six required Senning revision. The introduction of the Shumacker modification of the Senning has generated stable pulmonary venous pathways. Six patients developed postoperative iatrogenic atrioventricular block dependent on a permanent pacemaker.

**Conclusions:** Anatomic correction is a surgical challenge. It provides excellent functional outcomes in survivors with a significant need for reoperation and a definite risk of death or transplantation. Normal LV function should be expected in most patients. LV dysfunction was not linked to PAB or pacemaker requirement but surgery without LV training had better long-term LV function. The Shumacker modification provided stable venous pathways. Iatrogenic atrioventricular block remains a challenge. (J Thorac Cardiovasc Surg 2017;154:256-65)



Senning modification according to Shumacker creating large compliance venous chambers.

**Central Message**

Anatomic correction for congenitally corrected transposition of the great arteries is a complex surgery. It provides excellent functional outcomes in survivors.

**Perspective**

Understanding the long-term results of anatomic correction for congenitally corrected transposition of the great arteries allows for improvement in the consideration of timing, strategy, and technical details of the surgery in these patients. This will subsequently improve patient outcomes in our center and others.

See Editorial Commentary page 266.

Symptoms or evidence of declining cardiac function in patients with congenitally corrected transposition of the great arteries (ccTGA) require surgical management to control the evolution of cardiac failure. The results of physiologic correction maintaining a systemic right ventricle (RV) have been poor,[1-3] whereas anatomic correction surgery restores the morphologic left ventricle (LV) and mitral valve (MV) into their systemic role.[4] The literature

Scanning this QR code will take you to the supplemental tables, figures, and video for this article.

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*Read at the 96th Annual Meeting of The American Association for Thoracic Surgery, Baltimore, Maryland, May 14-18, 2016.*

Received for publication May 25, 2016; revisions received Jan 19, 2017; accepted for publication March 5, 2017; available ahead of print May 2, 2017.

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0022-5223/$36.00



http://dx.doi.org/10.1016/j.jtcvs.2017.03.072

**Abbreviations and Acronyms**

AR = aortic regurgitation
ccTGA = congenitally corrected transposition of the great arteries
CHB = complete heart block
LV = left ventricle
LVOT = left ventricular outflow tract
MR = mitral regurgitation
MV = mitral valve
neo-AV = neoaortic valve
PAB = pulmonary artery band
PPM = permanent pacemaker
RCH = Royal Children's Hospital
RV = right ventricle
TR = tricuspid regurgitation
TV = tricuspid valve
VSD = ventricular septal defect

surrounding this procedure is expanding. The procedure combines the correction of the atrioventricular discordance using an atrial switch (Senning procedure at The Royal Children's Hospital [RCH]), and simultaneously the correction of ventriculoarterial discordance. Thus, the combined techniques are the Senning-arterial switch or double-switch, Senning-Rastelli procedure, and Senning-Bex/Nikaidoh procedure.

This study aimed to contribute to the body of knowledge and to further refine the surgical indication, timing, and pitfalls of anatomic correction of ccTGA by reviewing our experience during the past 19 years.

## METHODS

### Study Population

This study was reviewed and approved by the RCH Human Research Ethics Committee (reference No. 34184A). The need for patient consent was waived due to the retrospective nature of the study.

ccTGA was defined as patients with discordant atrioventricular and ventriculoarterial connections. Morphologic diagnoses of patients were established by 2-dimensional echocardiography and confirmed at surgery. The hospital database was screened to identify patients with a diagnosis of ccTGA or synonymous terms. Of 84 children known to RCH with this diagnosis up to August 2015, 32 patients had undergone 1 of 3 anatomic correction procedures from 1996 onward at RCH and were included in this study (Figure E1). Twenty-seven patients with ccTGA had their decisive surgery during the same time span. There were 16 Fontan procedures for unbalanced ventricles, 7 Fontan procedures for unsuitable anatomy for repair, and 4 physiologic repairs.

### Follow-up

Preoperative, operative, and follow-up data on the patients were collected from hospital databases and from referring cardiologists between March and November 2015. One international patient was lost to follow-up.

An objective review of incomplete echocardiographic reports was obtained from a cardiologist at RCH. The severity of valvular regurgitation was graded from 0 (none), 1 (trivial), 2 (mild), 3 (moderate), to 4 (severe).[5] Subjective evaluation of LV function was utilized when no objective measure was recorded. For those studies where an objective measure of ejection fraction was available, we used the correlation of normal function for ejection fraction > 51%, mild dysfunction 41% to 50%, moderate dysfunction 31% to 40%, and severe dysfunction ≤ 30%.

### Statistical Analysis

All data were exported to and analyzed using IBM SPSS Statistics version 22.0 (IBM-SPSS Inc, Armonk, NY) and R version 3.1.3 (R Foundation for Statistical Computing, Vienna, Austria). Continuous variables were reported as median and range and/or interquartile range (IQR). Categorical variables were reported as counts and associated percentages. Analyses between categorical variables employed 2-sided Pearson $\chi^2$ tests. Competing risks survival methods were used to describe the cumulative incidence of the event of late reintervention and reoperation with death and heart transplantation as the competing risks. Kaplan-Meier curves were used to describe freedom from death or heart transplantation following anatomic correction. Cox proportional hazards models and likelihood ratio tests were used to assess the association of prior pulmonary artery band with the event of aortic valve surgery or neoaortic valve (neo-AV) regurgitation moderate or greater, and the effect of age at surgery (in years), permanent pacemaker insertion (time-varying covariate), and prior pulmonary artery band (PAB) with the event of reduced LV function at follow-up postanatomic correction. Event rates at 1, 5, and 10 years were reported with corresponding 95% confidence intervals (CIs). No multivariate analysis was performed due to the small numbers of events.

### Definitions

Early death, reintervention, or reoperation was defined as occurring before hospital discharge or within 30 days of surgery. Late death, reintervention, or reoperation was defined as occurring after discharge and more than 30 days after surgery.

Permanent pacemaker (PPM) placement is the first instance of PPM placement. PPM revision is a change in pacemaker leads or generator box at any time after initial PPM placement.

## RESULTS

### Morphologic Data

There were 18 male patients (56%). Characteristics of the patients are detailed in Table 1. Twenty-nine patients (91%) had situs solitus, 2 (6%) patients had situs inversus, and 1 had left isomerism (3%). This latter patient was kept in the series because he underwent an anatomic correction procedure. Coronary anatomy was confirmed as 1LADCX2R in 19 (59%) patients.

A ventricular septal defect (VSD) was present in 25 patients (78%); 13 patients had associated pulmonary or subpulmonary stenosis, and 12 patients had no pulmonary circulation protection. Eight (25%) patients had features of Ebstein's anomaly of the tricuspid valve (TV).

Nine patients (28%) had a conduction anomaly before corrective surgery. One had acquired complete heart block (CHB) following a palliative procedure performed outside the RCH (LV outflow tract [LVOT] resection), 3 had complete heart block, 4 had second-degree heart block, and 1 had first-degree heart block.

Preoperatively, 31 (97%) patients had normal LV function and 27 (84%) had normal RV function. Seven (22%) patients had moderate or greater tricuspid regurgitation (TR). No