Outcomes of definitive surgical repair for congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections: Risk analyses in 189 patients

Toshiharu Shin'oka, MD, PhD, Hiromi Kurosawa, MD, PhD, Yasuharu Imai, MD, PhD, Mitsuru Aoki, MD, PhD, Masakuni Ishiyama, MD, PhD, Takahiko Sakamoto, MD, PhD, Shinka Miyamoto, MD, Kyoko Hobo, MD, and Yuki Ichihara, MD

Dr Shin'oka

Supplemental material is available online.

From the Department of Cardiovascular Surgery, The Heart Institute of Japan, Tokyo Women's Medical University, Tokyo, Japan.

Read at the Eighty-sixth Annual Meeting of The American Association for Thoracic Surgery, Philadelphia, Pa, April 29–May 3, 2006

Received for publication July 25, 2006; revisions received Oct 25, 2006; accepted for publication Nov 3, 2006.

Address for reprints: Dr Toshiharu Shin'oka, Department of Cardiovascular Surgery, The Heart Institute of Japan, Tokyo Women's Medical University, 8-1 Kawada-cho, Shinjuku-ku, Tokyo 162-8666, Japan (E-mail: ssinoka@hij.twmu.ac.jp).

J Thorac Cardiovasc Surg 2007;133:1318-28

0022-5223/$32.00

doi:10.1016/j.jtcvs.2006.11.063

Objective: This study was undertaken to compare long-term results of various types of surgical repairs for either congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections, and to analyze the risk factors that affect early and late mortality and reintervention.

Methods: Between January 1972 and September 2005, a total of 189 patients (median age 8.3 years, range 2 months to 47 years old) with congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections underwent definitive repairs. The definitive repairs comprised a conventional repair (atrial septal defect, or ventricular septal defect closure with or without pulmonary stenosis release, or isolated tricuspid valve surgery) in 36 patients (group I), conventional Rastelli in 31 patients (group II), double-switch operation (atrial switch plus arterial switch) in 15 patients (group III), atrial switch plus intraventricular rerouting (with or without extracardiac conduits) in 69 patients (group IV), and a Fontan-type repair in 38 patients (group V). The mean follow-up period was 10.1 years. Hospitalization and late mortality and reoperation were indicated as events. Risk factors for these events were analyzed by logistic regression for hospital death and a Cox proportional hazards model for late events.

Results: The Kaplan–Meier survival including hospital and late mortality was 62.4% at 32 years in group I, 78.5% at 27 years in group II, 74.5% at 15 years in group III, 80% at 16 years in group IV, and 79.3% at 22 years in group V. The reoperation-free ratio was 64.2% in group I, 76.6% in group II, 84.4% in group III, 89.6% in group IV, and 91.3% in group V. Risk analyses showed that the risk for hospital death was preoperative in patients with more than moderate tricuspid regurgitation and a cardiopulmonary bypass time of more than 240 minutes. A risk for late mortality was the presence of tricuspid regurgitation. Risks for reoperation were preoperative cardiomegaly, preoperative tricuspid regurgitation of more than grade II, ventricular septal defect enlargement, and body weight less than 10 kg. Risks for pacemaker implantation, as indicated by multivariate analysis, were ventricular septal defect enlargement during operation and age less than 3 years.

Conclusions: There were no statistical differences between long-term survival rates of patients who underwent conventional surgical repair versus those of patients who underwent anatomic surgical repair. Results of conventional repair were satisfactory except in patients with significant tricuspid regurgitation. Results of anatomic repair were also satisfactory even for patients with significant tricuspid regurgitation, and therefore, anatomic repair should be the procedure of choice for those patients.

CHD

Abbreviations and Acronyms

ASD = atrial septal defect
AV = atrioventricular
CTR = cardiothoracic ratio
LD = late mortality
PA = pulmonary atresia
PM = pacemaker
PS = pulmonary stenosis
RA = right atrium
RV = right ventricular
TR = tricuspid regurgitation
TVR = valve replacement
VSD = ventricular septal defect

Congenitally corrected transposition of the great arteries (cc-TGA) or atrioventricular (AV) discordance with a double-outlet right ventricle (l-DORV) are characterized by AV and ventriculoarterial discordance.[1] These two entities were thought to differ only in the grade of overriding of the posterior pulmonary artery. When both septums in the heart are intact, the circulation is physiologically normal, with no shunt, no pressure load, and no cyanosis. However, the anatomic right ventricle and tricuspid valve must sustain a systemic pressure. Before 1983, in our institute, conventional repair was the procedure of choice. However, we found that, after conventional repair, some of our patients developed a systemic right ventricular (RV) dysfunction or tricuspid regurgitation (TR) over the long term. Because of the disappointing results with conventional repairs, the first patient with cc-TGA underwent a Mustard–Rastelli procedure that resulted in early mortality in 1983. In 1989, a second patient successfully survived a double-switch operation (DSO). Since then, anatomic repair has become the procedure of choice in our institute (Figure E1). The purpose of this study was to compare long-term outcomes after conventional versus anatomic repair for cc-TGA/ l-DORV. We also reviewed long-term results after a Fontan-type procedure for cc-TGA/ l-DORV, although this comparison is only for reference because the biventricular repair was not feasible in this group. In addition, we had 6 patients with anatomically corrected malposition who underwent definitive correction using the anatomic left ventricle as a systemic ventricle without any mortality. These patients were excluded in this study because systemic ventricles are the anatomic left ventricles after corrective surgery without any switch procedures.

Materials and Methods

Definitions of Terms

Terms were defined as in Ilbawi's descriptions.[2] Briefly, the terms "right ventricle" and "left ventricle" describe ventricular morphology rather than spatial orientation. "Tricuspid valve" refers to the AV valve associated with the anatomic right ventricle regardless of its location. Regarding the surgical procedure, "conventional repair" includes a conventional repair and conventional Rastelli procedure using the anatomic right ventricle as a systemic ventricle, and "anatomical repair" indicates a DSO or Senning/Mustard plus Rastelli procedure in which the anatomic left ventricle functions as a systemic ventricle.

Patient Population and Data Acquisition

One hundred eighty-nine patients with segmental anatomy [SLL] (n = 147) or [IDD] (n = 42) who underwent definitive surgical repair at Tokyo Women's Medical University between January 1972 and September 2005 were identified from the database of the Departments of Cardiovascular Surgery and Pediatric Cardiology. Medical records, preoperative and postoperative echocardiograph data, and cardiac catheterization data and operative notes were reviewed. Long-term follow-up was obtained by review of patient records and outpatient clinic notes. Institutional Review Board approval was obtained prior to the initiation of these retrospective analyses.

Diagnosis

Patient diagnoses were categorized in the following 6 groups according to the associated cardiac lesions (Table 1).

1. Hemodynamically significant ventricular septal defect (VSD) alone
2. Hemodynamically significant VSD with pulmonary stenosis (PS)
3. Hemodynamically significant VSD with pulmonary atresia
4. Hemodynamically significant VSD with TR
5. TR alone
6. Others (including atrial septal defect [ASD] alone, PS alone, atrioventricular septal defect [AVSD], etc)

If both of the great arteries arose from the anatomic right ventricle for more than 150% or a truncal atresia of the pulmonary artery existed, a diagnosis of DORV was given. AV valve regurgitation was judged as none, I, II, III, and IV by Seller's classification. More than grade II regurgitation was defined as significant.

Surgical Procedures

The surgical procedures comprised a conventional repair (ASD or VSD closure ± relief of PS, or isolated tricuspid valve surgery) in 36 patients (group I: conventional), conventional Rastelli in 31 patients (group II: conventional Rastelli), DSO (atrial switch plus arterial switch) in 15 patients (group III: DSO), atrial switch plus intraventricular rerouting (±extracardiac conduits) in 69 patients (group IV: M/S-Rastelli), and a Fontan-type repair in 38 patients (group V: Fontan).

Group I: Conventional Repair (1972–2003; n = 36)

Two patients had a complete form of AV septal defect; both defects were repaired by a conventional method, with 1 late death. Twelve of 36 patients required surgery for TR: valve replacement (TVR) in 8 patients and valve repairs in 4 patients. A mechanical valve was used in 6 patients, and a bioprosthetic valve was used in 2 patients. One patient with TVR had concomitant left ventricular (LV) assist device implantation with later heart transplantation. Three of 8 patients with TVR underwent PA banding for left ventricle training and failed.