Ideal timing of surgical repair of isolated complete atrioventricular septal defect

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Abstract

The ideal timing and optimal management of surgical repair for isolated complete atrioventricular septal defect (CAVSD) still remains controversial. To determine outcomes after the surgical repair of CAVSD, we reviewed 100 consecutive patients who underwent complete repair at our institute between January 1992 and August 2003. Among these 100 patients, 52 were female and 73 had Down’s syndrome. Twelve had received preceding pulmonary artery banding. A two-patch repair was employed in all cases. The patients’ median age and weight were 4.5 (1.2–48) months and 4.7 (2.5–12.5) kg, respectively. The mean stays in the intensive care unit and in the hospital were 5.3 ± 3.8 and 25.4 ± 18.1 days, respectively. The median duration of mechanical ventilation was 11.6 h. There were two in-hospital deaths, in patients 5.2 and 5.9 months of age. Both had underlying Down’s syndrome, and significant pulmonary vascular obstructive disease (PVOD) was detected on postmortem specimen. The operative outcome of CAVSD was generally satisfactory. PVOD can progress rapidly from four to five months, especially with Down’s syndrome. Therefore, in order to avoid progression to irreversible PVOD, surgical intervention within four months of birth may be appropriate in such patients.

Keywords: Complete atrioventricular septal defect; Survival analysis; Pediatric cardiac surgery

1. Introduction

Half a century has passed since Lillehei and colleagues successfully corrected a complete atrioventricular septal defect (CAVSD) [1]. Improved surgical results after repair of CAVSD have been reported owing to refinements in surgical technique, improved myocardial protection, and a better understanding of the surgical anatomy [2–4]. However, the ideal timing for the repair and the optimal management strategy (primary or two-stage repair) remains controversial. Some authors advocate primary repair at the age of six months or earlier [5,6], which has also been our institutional policy.

In this study, we retrospectively reviewed our recent experience with the surgical repair of isolated CAVSD with a focus on the ideal timing and management of surgery.

2. Material and methods

2.1. Patients characteristics

Between January 1992 and August 2003, a total of 100 consecutive patients (48 males and 52 females) with isolated CAVSD underwent total repair at Sakakibara Heart Institute. Seventy-three of these patients had Down’s syndrome. Those with other major associated cardiovascular anomalies – such as tetralogy of Fallot, double-outlet right ventricle, transposition of the great arteries, total anomalous pulmonary venous connection, and coarctation of the aorta – were excluded. Forty-five patients had Rastelli type A anatomy; the other 55 patients had type C anatomy. The patients’ median age was 4.5 (1.2–48) months, and their median body weight was 4.7 (2.5–12.5) kg. All patients received diagnostic catheterization, which provided data permitting the calculation of pulmonary vascular resistance (PVR). Our gross institutional criteria for the performance of primary complete repair are as follows: (1) an ample-sized mural leaflet, (2) normal chordal attachment, (3) two separate papillary muscles more than 1 cm apart, (4) absence of an accessory valve orifice, (5) presence of two balanced ventricles, and (6) PVR < 8 U/m² with or without oxygen administration.

For patients with PVR 8 U/m² that is unresponsive to oxygen inhalation, the initial approach includes banding of the pulmonary artery (PA) with or without lung biopsy. The histological staging of the specimen is done microscopically. The progression of pulmonary vascular obstructive disease (PVOD) is assessed according to the thickness of the media in relation to the entire pulmonary vascular wall. Subsequent treatment options are decided according to the results of histological staging together with data from a repeated catheterization [7] (Fig. 1).

2.2. Surgical technique

Moderate hypothermic cardiopulmonary bypass (CPB) was used in all patients, with pharyngeal temperature reduced...
to 27 °C. Bypass flow was adjusted at 150 ml/kg/min. All patients received two-patch repair using either polytetrafluoroethylene, autologous pericardium, or glutaraldehyde-treated equine pericardium. The cleft of the left atrioventricular valve was completely closed in all patients.

2.3. Statistical analysis

The SPSS statistical software for Windows (Version 11.0, SPSS Inc., Chicago, IL) was used for data analysis. Actuarial and freedom from reoperation curves were constructed using the Kaplan–Meier method. The log-rank test was used to ascertain the difference between those who were above four months of age at the time of surgery and those who were below that age. Univariate analysis with overall survival was assessed on the basis of separate Cox regression models. Predictive factors for survival in all patients were age, weight, gender, PA banding (PAB), length of stay in the intensive care units (ICU) and hospital, ventilator support time, operative time, CPB time, Rastelli type, and myocardial ischemia time. Multivariable Cox regression models using a stepwise variable selection method was used to identify the factors most significantly associated with survival. A P-value < 0.05 was considered as statistically significant. Data are presented as mean value ± standard deviation or as median where indicated.

3. Results

Preoperative systolic pulmonary arterial pressure was 68.1 ± 18.7 mmHg. Twenty-four patients received postoperative catheterization. The postoperative systolic pulmonary arterial pressure was 36.5 ± 11.2 mmHg.

Based on our diagnostic criteria, complete repair was performed as a first-time procedure in 87 patients. PAB was performed in 12 patients, and ligation of a patent ductus arteriosus (PDA) was done in the remaining patient. The PDA ligation and initial PAB procedures were done in referring hospitals. In seven patients, the reason for performing PAB in our institute was primarily the anatomical problem posed by the atrioventricular valve and its papillary muscles. The mean age at PAB at our institute was 3.2 ± 1.9 months. After PAB and PDA ligation, all 13 patients underwent biventricular repair at 17.5 ± 12.5 months of age.

The mean CPB and myocardial ischemia times were 123.6 ± 31.7 and 87.7 ± 23.6 min, respectively. The mean stay in the ICU and the hospital were 5.3 ± 3.8 and 25.4 ± 18.1 days, respectively. The median duration of mechanical ventilation was 11.6 h.

Postoperative complications were seen in 11 patients; two had wound infection and dehiscence, three had atrial flutter, and six required prolonged mechanical ventilatory support (more than seven days). With regard to the function of the atrioventricular valve, 12 patients had mild to moderate mitral valvular regurgitation. Of these, two had severe regurgitation requiring reoperation, while six had mild to moderate tricuspid regurgitation and one had severe tricuspid regurgitation.

Three patients underwent reoperation within 30 days because of residual shunt via a ventricular septal defect, postoperative hemolytic urine, and severe mitral regurgitation, respectively. The other four patients underwent reoperation 11 months after complete repair. Two had mitral stenosis and regurgitation and received mitral valve replacement using a St. Jude medical prosthesis (18 mm) and an ATS prosthesis (18 mm), respectively. One of these patients underwent repair of a residual VSD and another had concomitant surgery to relieve left ventricular tract outflow obstruction.

The Kaplan–Meier estimated overall actuarial survival was 94.3 ± 5.7% at 12.5 years (Fig. 2). The actuarial survival rates between those who were above versus below four months of age at the time of operation were 9.20 ± 3.9% and 97.5 ± 2.4%, respectively (P = 0.29). Univariate analysis identified CPB time as a predictive factor for survival (P = 0.03). In the multivariate analysis, the same result was found (P = 0.03). All other factors failed to reach statistical significance. Overall freedom from reoperation was 92.0 ± 2.9% at 12.5 years (Fig. 3). The incidence of reoperation was concentrated within two years after complete repair. No patient required reoperation after this period. Two patients, both with Down’s syndrome, died in the early postoperative period. The postmortem pathological examination of the PA in both patients revealed that the media of the small PAs was extremely hypertrophic. Peripheral PAs < 0.2 mm in diameter were almost entirely obstructed due to a thickened media and fibrous hypertrophy of the
internal membrane. The catheterization data confirmed the feasibility of surgery; however, findings of irreversible PVOD were detected in the postmortem lung specimen. We speculate that, in this specific patient group with Down’s syndrome, it is possible for PVOD to progress rapidly between four and five months of age.

Late mortality was seen in three patients with Down’s syndrome. The ages of these patients at operation were 3.3, 4.5, and 10.5 months, respectively. One patient died of a respiratory infection 47 months after surgery. Another died of cardiac insufficiency with residual pulmonary hypertension at three years. In the remaining patient, death occurred at 10.5 months, but the cause was not determined.

4. Discussion

The outcome after surgery for CAVSD has improved owing to a better understanding of the disease physiology together with improved surgical techniques. Today, many surgeons prefer early primary repair without intervening palliative procedures. Reported mortalities resulting from this practice are generally lower than those reported after two-stage repair. Surgery before six months of age is usually recommended [5,6]. However, there is little information on the ideal timing of repair. Irreversible PVOD can develop before these patients reach six months of age, especially in the presence of Down’s syndrome [8]. Such patients may benefit from earlier repair.

Of note, one patient who received complete repair before the age of four months died, and four additional deaths occurred among those who underwent surgery after four months of age. The cause of death in two of the latter four patients was undoubtedly related to PVOD. These patients received cardiac catheterization at 4.0 and 4.1 months of age. However, in both of these cases, there was an interval of more than one month before surgery took place. This suggests that the PVOD can progress rapidly between four and five months. Yamaki and colleagues indicate that PVOD in patients with complete AVSD and Down’s syndrome may progress more rapidly than it does in non-Down’s patients [8]. Therefore, in order to avoid progression to irreversible PVOD, surgical intervention within four months of birth may be appropriate in Down’s patients.

We also think that primary repair, rather than two-stage repair, is always to be preferred. PAB, however, may be the only option for patients in whom complete repair is not feasible. In this context, tight banding can cause severe pulmonary stenosis with myocardial hypertrophy, and loose banding can result in irreversible PVOD. Yamaki and colleagues have reported a patient with VSD who died of PVOD after PAB and subsequent total correction [9]. Therefore, patients who have received PAB should be followed strictly and carefully. In our institute, PAB is performed when the catheterization date does not meet the criteria outlined by Yamaki and coworkers. The later decision to perform a complete repair is guided by repeat catheterization after PAB and/or lung biopsy. The accurate interpretation of data is especially important, because PVOD is the main cause of mortality after the repair of CAVSD.

Univariate and multivariate analyses identified the CPB time as a predictive risk factor for postoperative mortality. The CPB generally promotes an inflammatory reaction; therefore, an effort to minimize CPB time is another key to the reduction of postoperative mortality.

In conclusion, the results of our surgical repair of isolated CAVSD were generally satisfactory. We believe that it is safe to perform a complete repair within four months of age in order to avoid progression to irreversible PVOD. To achieve a satisfactory outcome, accurate diagnostic evaluations consisting of echocardiography and catheterization are warranted.

References