

Long-term outcomes after surgical repair of complete atrioventricular septal defect

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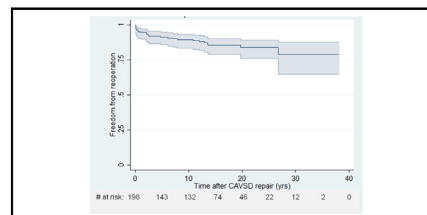
ABSTRACT

Objective: Survival after surgical repair for complete atrioventricular septal defect (CAVSD) has improved, but patients are at risk for reoperation to address left atrioventricular valve regurgitation and left ventricular outflow tract obstruction. We examined the long-term survival, need for reoperation, and associated risk factors after CAVSD repair at our institution.

Methods: Between 1974 and 2000, a total of 198 patients underwent surgical repair for CAVSD. Of these, 178 patients survived to hospital discharge, of whom 153 (86%) had available follow-up data at a median postoperative time point of 17.2 years (range: 2 months to 38.1 years).

Results: Overall perioperative mortality was 10.1%, with a significant decrease to 2.9% in the late surgical era: 1991 to 2000 ($P < .001$). The overall estimated survival for the entire cohort was 85% at 10 years, 82% at 20 years, and 71% at 30 years after initial CAVSD repair. Requiring a reoperation after initial CAVSD repair was a risk factor for late mortality ($P = .04$). The estimated freedom from reoperation was 88% at 10 years, 83% at 20 years, and 78% at 30 years after initial CAVSD repair. Indications for reoperation included left atrioventricular valve regurgitation in 14 patients (7.1%) and left ventricular outflow obstruction in 7 patients (3.5%).

Conclusions: Long-term survival after repair of CAVSD remains good. However, the need for reoperation is common and affects long-term survival after CAVSD repair. (J Thorac Cardiovasc Surg 2015;150:369-74)



Estimated freedom from reoperation after CAVSD repair.

Central Message

Reoperation after surgical repair for complete atrioventricular septal defect affects long-term morbidity and mortality.

Perspective

With a median follow-up of >17 years after repair of complete atrioventricular septal defect, this study provides outcomes data, to assist providers in caring for this growing population who survive into adulthood. Although survival has improved, risk for reoperation remains. These findings can help providers determine the need for and frequency of cardiology follow-up and testing.

See Editorial Commentary page 375.

The first successful surgical repair of a complete atrioventricular septal defect (CAVSD) was performed by C. Walton Lillehei in 1955.¹ Since that time, advances in surgical and medical management have resulted in decreased perioperative mortality and improved early survival.²⁻⁴ Previous studies^{5,6} have demonstrated a 15-year survival of 86% to 89%. However, despite improved survival, several single-center studies report that

up to 10% of patients, after initial CAVSD repair, may require reoperation within 10 to 15 years, to address left atrioventricular (AV) valve regurgitation and left ventricular outflow tract obstruction.^{3,5-8} Given that survivors of CAVSD repair are now presenting to the Adult Congenital Heart Disease Program in their third and fourth decades of life, we sought to determine the long-term survival and need for reoperation in patients after CAVSD repair, at our institution.

METHODS

Patient Population

A total of 198 patients who had undergone complete surgical repair for CAVSD between the years 1974 and 2000 were identified from the Children's Hospital of Wisconsin surgical database. Patients with an associated diagnosis of tetralogy of Fallot, defined as subvalvular and/or valvular pulmonary stenosis that required intervention at the time of CAVSD repair, were excluded from this total. Patients with partial AV septal defect (ostium primum atrial septal defect), intermediate-type AV

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Abbreviations and Acronyms

AV = atrioventricular
CAVSD = complete atrioventricular septal defect

septal defect (a common AV valve or 2 separate AV valve orifices, but restrictive or absent ventricular septal defect), or unbalanced forms of AV septal defect who underwent single-ventricle palliation were excluded. The protocol was approved by the Children's Hospital of Wisconsin Institutional Review Board.

Surgical Era and Technique

The 198 patients were divided into 2 groups, to determine the impact of surgical era on long-term outcomes. The early surgical era included those patients who underwent repair from 1974 to 1990 ($n = 93$; 47%); the late surgical era included patients who underwent repair from 1991 to 2000 ($n = 105$; 53%). Surgical repair of CAVSD was performed with either the classic 1-patch technique or the 2-patch technique, along with closure of the left AV valve cleft, as previously described.⁹ Briefly, the classic 1-patch technique was performed by dividing the common superior and inferior bridging leaflets, leaving slightly more tissue on the left side of the common AV valve. A single Dacron patch was sutured onto the right side of the ventricular septum, to close the septal defect, and the AV valve components were attached to the patch using felted mattress sutures, along with closure of the cleft of the left AV valve leaflet.

The 2-patch technique was performed by first closing the ventricular septal defect with a Dacron patch. The ventricular septal defect patch was positioned under the bridging leaflets and secured to the right side of the ventricular septum. Sutures were passed through the superior edge of the patch and through the bridging leaflets at the point of division between the right and left components of the common AV valve. These same sutures were passed through a second atrial septal defect patch and tied. The cleft of the newly constructed left AV valve was closed with interrupted sutures before completion of the atrial septal defect closure. Most patients in the early surgical era underwent repair with the classic 1-patch technique (79.6%), compared with the 2-patch technique (20.4%). In the late surgical era, most patients underwent the 2-patch technique (98%). No patient underwent repair with the modified 1-patch technique.

Study Endpoints

Primary outcomes for the study included survival, and freedom from reoperation after initial CAVSD repair. Early death was defined as death within 30 days after CAVSD repair or before hospital discharge. Late death was defined as death beyond 30 days after CAVSD repair and after hospital discharge. Hospital survivor was defined as surviving beyond 30 days after CAVSD repair, and after hospital discharge. Reoperations included any cardiac surgery after CAVSD repair. Interventional cardiac catheterizations and permanent pacemaker placements performed during the study period were not included as reoperations but are discussed separately.

Data Collection and Follow-up

A retrospective analysis of medical records was performed for all 198 patients, to identify operative and long-term mortality and need for reoperation after CAVSD repair, as well as risk factors for mortality and reoperation, including surgical technique, surgical era, age at repair, and associated diagnoses. For the 178 hospital survivors, a total of 153 patients (86.0%) had available and current data within the past 5 years that were obtained from either the medical record ($n = 132$) or a medical questionnaire that was sent to and returned by the patient's primary care physician inquiring about recent clinical status ($n = 21$). The mean

follow-up for hospital survivors was 17.42 ± 7.8 years (median = 17.2 years; range = 2 months to 38.1 years).

Statistical Analysis

Continuous variables are reported as mean \pm SD, or median with range, and categoric variables are presented as frequencies and percentage of total. Groups were compared using χ^2 analysis or the Fisher exact test for categoric variables and the Wilcoxon rank sum test for continuous variables. Cox proportional hazards models were used to evaluate the risk factors for mortality and need for reoperation. Risk factors for left AV valve reoperation and left ventricular outflow tract obstruction surgery were evaluated separately because these risks may occur at different time points. For these models, log transformation of age at initial repair was required, and interactions between variables were evaluated. Estimates of overall survival and freedom from late reoperation were calculated using the Kaplan-Meier method and compared, when necessary, using the log-rank test. Data were analyzed using StataIC 13 (Stata Corporation, College Station, Tex).

RESULTS**Early Operative Patient Characteristics**

The patient characteristics for the 198 patients that underwent complete surgical repair for CAVSD are summarized in Table 1. The mean age at the time of repair was 1.36 ± 1.9 years (median = 8.1 months; range = 1 month to 15 years). The mean age decreased, from 1.96 years in the early surgical era to 9.9 months in the late surgical era.

Associated diagnoses included Down syndrome in 156 patients (78.8%), coarctation of aorta in 6 (3.0%), and left ventricular outflow tract obstruction in 2 (1.0%). A total of 32 patients (16.2%) underwent palliation with a pulmonary artery band before complete repair of CAVSD.

Mortality

A total of 20 early operative deaths occurred, defined as death within 30 days after CAVSD repair or before hospital discharge, resulting in an overall early mortality of 10.1%. The early mortality decreased significantly from 18.2% (17 of 93 patients) in the early surgical era to 2.9% (3 of 105 patients) in the late surgical era ($P < .001$).

Among the 178 hospital survivors, 15 late deaths occurred after initial CAVSD repair. The cause of late death was cardiac related in 9 patients. Causes of cardiac-related deaths included: complications related to congestive heart failure in 1 patient who had a large residual ventricular septal defect who developed pneumonia and respiratory failure; 1 patient with severe left AV valve regurgitation who died in the postoperative period after left AV valve replacement at age 2 months; 1 patient with late diagnosis of coarctation of aorta who died in the postoperative period after surgical aortic arch repair; 2 patients with dilated cardiomyopathy of unknown etiology that may have been related to a history of complete heart block and/or the need for chronic ventricular pacing; and 3 patients with sudden death. The 3 patients with sudden deaths all had

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