More Than 30 Years' Experience With Surgical Correction of Atrioventricular Septal Defects

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Background. **Outcome of surgical correction of atrioventricular septal defects (AVSD) still varies despite enhanced results. We reviewed our 30-year experience with AVSD repair and identified risk factors for mortality and reoperation.**

Methods. **Between 1975 and 2006, 312 patients underwent surgery for complete AVSD (n = 209; 67.0%), partial AVSD (n = 76; 24.4%), or intermediate AVSD (n = 27; 8.6%). Mean age was 2.4 ± 3.9 years; 142 patients (45.5%) were younger than 6 months. Follow-up was 99.0% complete.**

Results. **There were 26 in-hospital deaths (8.3%) and 6 late deaths (2.1% of 283). Estimated overall survival for the total study population was 91.3%, 90.6%, and 88.6% at 1, 5, and 15 years, respectively. In the multivariable logistic regression analysis, surgical era 1975 to 1995 ($p < 0.001$) and younger age ($p = 0.004$) were found to be independent risk factors for early mortality, whereas preoperative AV valve insufficiency showed a tendency toward statistical significance ($p = 0.052$). Of the hospital survivors, 43 patients required a late reoperation. Estimated freedom from late reoperation was 96.4%, 89.3%, and 81.8% at 1, 5, and 15 years, respectively. Multivariable Cox regression analysis showed associated cardiovascular anomalies ($p < 0.001$), left AV valve dysplasia ($p < 0.001$), and absence of cleft closure ($p = 0.003$) to be independent risk factors for late reoperation.**

Conclusions. **AVSD repair can be accomplished with good long-term results. Early surgical era, associated cardiovascular anomalies, left AV valve dysplasia, and absence of cleft closure negatively influence survival and risk of reoperation.**

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Atrioventricular septal defect (AVSD) includes complete, partial, and intermediate AVSD. In complete AVSD (c-AVSD), there is a common AV valve for both ventricles and interatrial and interventricular communication. In partial AVSD (p-AVSD), also referred to as primum type atrial septal defect (ASD I), there are separate right and left AV valve orifices, and interventricular communication is lacking. An intermediate form of AVSD (i-AVSD) is defined as having a "scooped out" interventricular septum with the AV valves being connected to the top of the septum by fibrous tissue "curtains" and tendinous chordae, consequently resulting in a small or absent VSD component.

Most investigations that evaluate outcome of surgical correction of AVSD focus on either c-AVSD or p-AVSD [1-6]. In this study, we evaluated whether type of AVSD (c-AVSD, p-AVSD, or i-AVSD), among other factors, is a risk factor for mortality and reoperation after surgical repair. In addition, we studied whether risk factors for mortality and reoperation differed for the three AVSD types. Hereto, we reviewed our institution's 30-year experience with surgical repair of AVSD.

Patients and Methods

Patient Population

Between January 1975 and May 2006, 312 consecutive patients underwent surgical correction of AVSD at our institution. Complete AVSD was observed in 209 patients (67.0%), p-AVSD in 76 (24.4%), and i-AVSD in 27 (8.6%). Patients with associated cardiovascular anomalies, namely, tetralogy of Fallot (TOF [n = 24; 7.7%]), presence of an accessory orifice within the left-sided AV valve (double orifice [DO] left AV valve; n = 21; 6.7%), coarctation of aorta (n = 11; 3.5%), and left ventricular outflow tract obstruction (n = 2; 0.6%), were included. Three patients (1.0%) were diagnosed with combined DO left AV valve–coarctation of aorta. All patients with unbalanced forms of AVSD or common AV valve malalignment were excluded, as were patients with other associated cardiovascular anomalies. Secundum type ASD and

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Abbreviations and Acronyms

AV = atrioventricular
AVSD = atrioventricular septal defect
c-AVSD = complete atrioventricular septal defect
CI = confidence interval
DO = double orifice
HR = hazard ratio
i-AVSD = intermediate atrioventricular septal defect
OR = odds ratio
p-AVSD = partial atrioventricular septal defect
TOF = tetralogy of Fallot

patent ductus arteriosus were not marked as associated cardiovascular anomalies. The study was approved with waiver of consent by the Ethics Committee of the institution.

Surgical Technique

Six pediatric cardiac surgeons performed 297 of the 312 procedures, with very little variation in surgical technique over time. Some older patients with p-AVSD were operated on by surgeons for adult patients. All operations were performed with cardiopulmonary bypass and moderate hypothermia. St. Thomas cardioplegia solution administered every 30 minutes was used throughout the years. Intraoperative transesophageal echocardiography both before and after repair was routinely used since 1999.

Atrioventricular Septal Defects

In patients with c-AVSD, the septal defects were all closed using a two-patch technique but with different patch materials (autologous or heterologous pericardium, or Dacron [C.R. Bard, Haverhill, PA]). A one-patch technique was used in all p-AVSD patients. In patients with i-AVSD, a two-patch technique was applied in 9 patients, and a one-patch technique served for repair in the other 18 patients. The cleft was closed by interrupted sutures to such an extent that regurgitation was managed optimally without creating a valvular stenosis. Cleft closure was performed in 180 of 209 c-AVSD patients (86.1%), in 42 of 76 p-AVSD patients (55.3%), and in 19 of 27 i-AVSD patients (70.4%).

Associated Cardiovascular Anomalies

Tetralogy of Fallot, DO left AV valve, coarctation of aorta, and left ventricular outflow tract obstruction were all simultaneously repaired with the AVSD. In the 24 patients with c-AVSD-TOF, right ventricular outflow tract enlargement was performed by a transatrial, transpulmonary approach. A transannular patch was used in 20 of 24 patients. The cleft was closed in 22 of 24 patients [7]. In the 21 patients with DO left AV valve, the accessory orifice was repaired in 14 patients with suture (n = 9), patch (n = 3), or resection of the tissue bridge and division of the papillary muscle of the accessory orifice (n = 2) [8].

Data Collection and Follow-Up

Patient data were obtained by reviewing both inpatient and outpatient medical records, including surgeon's notes, operative reports, hospital charts, and cardiac catheterization and echocardiographic reports. The majority of the patients underwent routine follow-up at our Department of Pediatric Cardiology. The closing interval for follow-up was February 2006 to December 2006. Follow-up data were complete for 283 of 286 hospital survivors (99.0%). For 7 patients, follow-up information was obtained by a telephone call to the patient's family or physician, or both. Three patients with c-AVSD were lost to follow-up. The median follow-up period was 10.7 years (range, 0.4 to 29.3). For c-AVSD patients (n = 184), median follow-up was 9.9 years (range, 0.4 to 28.9); for p-AVSD patients (n = 73), it was 15.0 years (range, 1.9 to 29.3); and for i-AVSD patients (n = 26), it was 6.2 years (range, 1.9 to 18.7).

Definitions and Endpoints

Left AV valve dysplasia was defined as a left AV valve with stiffened, thickened, and myxoid degenerated valve tissue and curling-in of the leaflets. Endpoints in the study were in-hospital and late mortality and early and late reoperation. In-hospital mortality and early reoperation were defined as death or reoperation before hospital discharge or within 30 days. Late mortality and reoperation were defined as all-cause death or reoperation more than 30 days after the surgical correction for AVSD. The AVSD repairs were divided into two surgical eras to assess the impact of surgical era on mortality and reoperation: 1975 to 1995, and 1996 to 2006. To assess the impact of concomitant procedures due to associated cardiovascular anomalies, associated cardiovascular anomalies were defined as absent or present because of the relatively small patient numbers for these anomalies in the three AVSD groups.

Statistical Analysis

Estimates of overall survival and freedom from late reoperation were obtained by means of the Kaplan-Meier method, with differences among the three AVSD groups being tested by the log rank test. Binary logistic regression analysis was used to examine the relationship between potential risk factors and in-hospital mortality. Separate logistic regression analyses were used first to assess the individual impact of each variable. Subsequently, a logistic regression analysis, using all variables in a stepwise method was performed to identify independent risk factors. Cox proportional hazard models (for each variable separately and for all variables combined in a stepwise method) were performed to examine predictors for late reoperation. Potential risk factors tested in relation to in-hospital mortality and late reoperation were surgical era, type of AVSD, age, sex, Down syndrome, associated cardiovascular anomalies, left AV valve dysplasia, preoperative left AV valve insufficiency, and cleft closure. Probabilities used for entrance and removal in the stepwise method were 0.05 and 0.10, respectively. A p value of less than 0.05 (two-sided) was