Contemporary outcomes of complete atrioventricular septal defect repair: Analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database

James D. St. Louis, MD,[a] Upinder Jodhka, MD, MHS,[b] Jeffrey P. Jacobs, MD,[c] Xia He, MS,[d] Kevin D. Hill, MD,[d] Sara K. Pasquali, MD, MHS,[e] and Marshall L. Jacobs, MD[c]

Objective: Contemporary outcomes data for complete atrioventricular septal defect (CAVSD) repair are limited. We sought to describe early outcomes of CAVSD repair across a large multicenter cohort, and explore potential associations with patient characteristics, including age, weight, and genetic syndromes.

Methods: Patients in the Society of Thoracic Surgeons Congenital Heart Surgery Database having repair of CAVSD (2008-2011) were included. Preoperative, operative, and outcomes data were described. Univariate associations between patient factors and outcomes were described.

Results: Of 2399 patients (101 centers), 78.4% had Down syndrome. Median age at surgery was 4.6 months (interquartile range, 3.5-6.1 months), with 11.8% (n = 284) aged ≤2.5 months. Median weight at surgery was 5.0 kg (interquartile range, 4.3-5.8 kg) with 6.3% (n = 151) < 3.5 kg. Pulmonary artery band removal at CAVSD repair was performed in 122 patients (4.6%). Major complications occurred in 9.8%, including permanent pacemaker implantation in 2.7%. Median postoperative length of stay (PLOS) was 8 days (interquartile range, 5-14 days). Overall hospital mortality was 3.0%. Weight < 3.5 kg and age ≤ 2.5 months were associated with higher mortality, longer PLOS, and increased frequency of major complications. Patients with Down syndrome had lower rates of mortality and morbidities than other patients; PLOS was similar.

Conclusions: In a contemporary multicenter cohort, most patients with CAVSD have repair early in the first year of life. Prior pulmonary artery band is rare. Hospital mortality is generally low, although patients at extremes of low weight and younger age have worse outcomes. Mortality and major complication rates are lower in patients with Down syndrome. (J Thorac Cardiovasc Surg 2014;148:2526-31)

CHD

The natural history of complete atrioventricular septal defect (CAVSD) includes premature death due to complications of congestive heart failure and/or pulmonary artery hypertension. Repair during infancy is recommended for all patients. Outcomes following surgical repair have improved over several decades due to refinements of technique and postoperative management.[1-7] The age for elective repair has steadily declined, from as late as 1 year a few decades ago to 3 to 6 months at most centers today.[6,8-12] Early repair is intended to minimize the risk of premature death or pulmonary vascular obstructive disease. There are, however, some patients with medically refractory congestive heart failure for whom repair even earlier in infancy must be considered, because the risk of adverse events during prolonged supportive medical therapy is substantial. Single-stage repair is generally preferred, but occasionally palliation with pulmonary artery banding (PAB) is considered.

Although the benefits of surgical repair before age 6 months are now generally acknowledged, optimal timing is still debated and is unlikely to be the same for all patients. A recent study by Atz and the Pediatric Heart Network (PHN) Investigators[9] reported outcomes of 120 patients who underwent repair of CAVSD at 7 centers. Adverse outcomes were negatively correlated with age at repair from birth to age 2.5 months, but did not vary by age beyond 2.5 months. There is a paucity of other multicenter studies addressing this issue.

CAVSD is frequently associated with Down syndrome.[10] Associations between Down syndrome and contemporary outcomes remain to be established. Infants with Down syndrome and left-to-right shunts have long been believed to be susceptible to pulmonary vascular reactivity and respiratory complications. Despite these risks, our previous analysis of

From the Division of Pediatric Cardiac Surgery,[a] Department of Surgery, and Department of Pediatrics,[b] University of Minnesota, Minneapolis, Minn; Division of Cardiac Surgery,[c] Department of Surgery, Johns Hopkins School of Medicine, Baltimore, Md; Duke Clinical Research Institute,[d] Duke University School of Medicine, Durham, NC; and Department of Pediatrics and Communicable Diseases,[e] University of Michigan, Ann Arbor, Mich.

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Address for reprints: James D. St. Louis, MD, East Building, 12th Fl, 2450 Riverside Ave, Minneapolis, MN 55454 (E-mail: stlou012@umn.edu).

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Abbreviations and Acronyms

CAVSD = complete atrioventricular septal defect
IQR = interquartile range
PAB = pulmonary artery banding
PHN = Pediatric Heart Network
PLOS = postoperative length of stay
STS-CHSD = Society of Thoracic Surgeons Congenital Heart Surgery Database

multi-institutional data from the Society of Thoracic Surgeons Congenital Heart Surgery Database (STS-CHSD) showed that mortality rates for patients with or without Down syndrome did not differ significantly across the spectrum of pediatric cardiac surgical procedures. Lengths of stay were prolonged for patients with Down syndrome undergoing some specific procedures, but CAVSD repair was not one of those.[13]

Apart from the PHN study,[9] there are no recent large multicenter studies pertaining to surgical outcomes among infants with CAVSD. The aim of our study is to provide a descriptive analysis of contemporary multicenter experience with repair of CAVSD using data from the STS-CHSD.

PATIENTS AND METHODS

Data Source

The STS-CHSD contains operative, perioperative, and outcomes data on more than 250,000 patients undergoing congenital heart surgery since 1998, and currently includes information from 108 participating hospitals. Data quality and reliability are assured through intrinsic verification of data and a formal process of site visits and data audits. The Duke Clinical Research Institute serves as the data warehouse and analytic center for all of the STS National Databases. This analysis was approved by the Duke University Institutional Review Board, and by the STS Access and Publications Committee. This study was not considered human subjects research by the Duke University Institutional Review Board in accordance with the common rule (45 CFR 46.102(f)).

Patient Population

All STS-CHSD records from 2008 to 2011 with the procedure code for CAVSD repair entered for the index operation were identified. Patients with concomitant diagnosis of tetralogy of Fallot, total anomalous pulmonary venous connection, double outlet right ventricle, or any form of single ventricle (including single ventricle unbalanced antrioventricular canal or single ventricle heterotaxy syndrome) were excluded. Cases were analyzed on the basis of the first CAVSD operation of each admission. Patients with missing or invalid data for key variables were excluded.

Data Collection

Patient characteristics included age, weight, weight-for-age z-score, gender, race/ethnicity, additional preoperative factors (as defined by the STS-CHSD), noncardiac congenital anatomic abnormalities, chromosomal abnormalities or syndromes, and prior cardiac surgical procedures. Procedural factors include cardiopulmonary bypass, crossclamp times, and concurrent procedures.

Outcomes

The primary outcome was discharge mortality. Postoperative length of stay (PLOS) was calculated from date of operation to date of hospital discharge. Both the occurrence of any postoperative complication collected in the STS-CHSD, as well as major complications were evaluated. The latter include renal failure requiring dialysis, neurologic deficit persisting at discharge, arrhythmia requiring permanent pacemaker, mechanical circulatory support, phrenic nerve injury/paralyzed diaphragm, and unplanned reoperation/reintervention.[14]

Analysis

Distributions of patient characteristics and outcomes were summarized in the overall sample, in patients with Down syndrome and those without, as well as in patient groups defined by age and weight at surgery. Specifically, the age groups ($\leq$2.5 and >2.5 months) were defined based on findings previously reported by the PHN investigators[9] and 3.5 kg was chosen as the threshold for low and high weight groups (<3.5 kg and $\geq$3.5 kg) after an initial exploration of discharge mortality by weight at surgery (Figure 1). In all groups, we reported the frequencies of categorical variables and the median values with interquartile ranges of continuous variables. Patient characteristics and outcomes were described between groups and compared using the χ^2 and Wilcoxon rank sum tests where appropriate. Due to the descriptive nature of this study, multivariable analyses were not undertaken. It is known that several key anatomic and physiologic variables that may influence outcome in this patient population are not currently collected in the STS-CHSD, thus limiting the utility and results of multivariable modeling to attempt to identify independent risk factors for adverse outcomes. All analyses were performed using SAS version 9.3 (SAS Institute, Inc, Cary, NC). A P value < .05 was considered statistically significant.

RESULTS

Patient Population

The cohort included 2399 patients over the 4 years of data collection (Table 1). Overall, 43% (n = 1040) of patients were male and 78% (n = 1882) had Down syndrome.

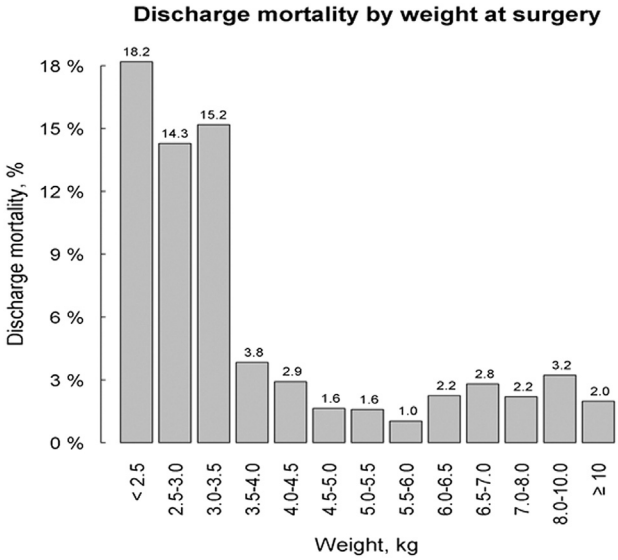

FIGURE 1. Discharge mortality by weight at surgery for the entire cohort of patients undergoing repair of complete atrioventricular septal defect during the period 2008 to 2011.