Surgical Interventions for Atrioventricular Septal Defect Subtypes: The Pediatric Heart Network Experience

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Background. **The influence of atrioventricular septal defect (AVSD) subtype on outcomes after repair is poorly understood.**

Methods. **Demographic, procedural, and outcome data were obtained 1 and 6 months after AVSD repair in an observational study conducted at 7 North American centers.**

Results. **The 215 AVSD patients were subtyped as 60 partial, 27 transitional, 120 complete, and 8 with canal-type VSD. Preoperatively, transitional patients had the highest prevalence of moderate or severe left atrioventricular valve regurgitation (LAVVR, $p = 0.01$). At repair, complete AVSD and canal-type VSD patients, both with the highest prevalence of trisomy 21 ($p < 0.001$), were younger ($p < 0.001$), had lower weight-for-age z scores ($p = 0.005$), and had more associated cardiac defects ($p < 0.001$). Annuloplasty was similar among subtypes ($p = 0.91$), with longer duration of ventilation and hospitalization for complete AVSD ($p < 0.001$). Independent predictors of moderate or severe LAVVR at the 6-month follow-up were older log(age) at repair ($p = 0.02$) but not annuloplasty, subtype, or center ($p > 0.4$). Weight-for-age z scores improved in all subtypes at the 6-month follow-up, and improvement was similar among subtypes ($p = 0.17$).**

Conclusions. **AVSD subtype was significantly associated with patient characteristics and clinical status before repair and influenced age at repair. Significant postoperative LAVVR is the most common sequela, with a similar prevalence across centers 6 months after the intervention. Annuloplasty failed to decrease the postoperative prevalence of moderate or severe LAVVR at 6 months. After accounting for age at repair, AVSD subtype was not associated with postoperative LAVVR severity or growth failure at 6 months. Further investigation is needed to determine if interventional strategies specific to AVSD subtype improve surgical outcomes.**

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Atrioventricular septal defects (AVSD) include a spectrum of abnormalities involving the atrial and ventricular septa and AV valves [1]. Techniques used for repairing AVSD subtypes vary among surgeons [2]. AVSD surgical outcomes have been the focus of benchmarking efforts and are used as a surrogate for institutional expertise in the management of congenital heart operations [3, 4, 5], but differences among the subtypes have not been systematically studied in the modern era. Using a multicenter observational database, we sought to compare patient characteristics, resource utilization, center differences, and outcomes among AVSD subtypes and to identify predictors of postoperative left AV valve regurgitation (LAVVR).

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Material and Methods

Between June 2004 and February 2006, all patients with AVSD undergoing biventricular repair at 7 North American centers were screened for potential enrollment into a drug trial designed to evaluate the effect of angiotensin-converting enzyme inhibitors on LAVVR and supported by the National Heart, Lung and Blood Institutes (NHLBI)–funded Pediatric Heart Network. Before the drug trial was initiated, echocardiographic and clinical data were collected 1 and 6 months postoperatively in an observational phase, as previously described [4, 5]. Briefly, data were collected prospectively collected for 178 patients who were participants in the planned drug trial and retrospectively for 37 screened patients from the same period with similar baseline characteristics who were not enrolled. Informed consent was obtained from parents/guardians for prospectively enrolled patients. A waiver of consent was obtained for retrospectively enrolled

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Abbreviation and Acronyms

ASD	= atrial septal defect
AVSD	= atrioventricular septal defect
CAVSD	= complete atrioventricular septal defect
CI	= confidence interval
LAVVR	= left atrioventricular valve regurgitation
LV	= left ventricular
NHLBI	= National Heart, Lung and Blood Institutes
OR	= odds ratio
PAVSD	= partial atrioventricular septal defect
TAVSD	= transitional atrioventricular septal defect
VSD	= ventricular septal defect

patients. The information in the database was obtained under the supervision of an independent Data and Safety Monitoring Board and approved by the center's Institutional Review Board (Clinical Trials.gov Identifier: NCT00113698).

Medical Records

Demographic data, including race/ethnicity, sex, presence of trisomy 21, and age and weight at operation and at the 6-month follow-up echocardiogram were recorded. Weight-for-age z scores were calculated using sex-specific reference values for trisomy 21 [6] and normal children [7], as appropriate. Days of mechanical ventilation, intensive care, and total hospitalization were recorded to estimate resource utilization.

Three investigators independently reviewed all operative reports for AVSD subtype, cleft closure, annuloplasty, and additional operations for associated defects. The Society of Thoracic Surgeons nomenclature was used to define AVSD subtypes [4, 5, 8]: partial AVSD (PAVSD) had a primum atrial septal defect (ASD) and cleft left AV valve (LAVV); transitional AVSD (TAVSD) had a primum ASD, restrictive inlet VSD and 2-orifice AVV; complete AVSD (CAVSD) had a primum ASD, inlet VSD, and single orifice AVV; and canal-type VSD had an inlet VSD, common AV junction, cleft LAVV, and no primum ASD.

Because the operative notes often contained insufficient detail regarding annuloplasty technique, annuloplasty was defined as any additional operation on the LAVV beyond cleft closure, such as annuloplasty along the lateral leaflet with absorbable suture, sutures at the superior and inferior bridging-lateral leaflet commissures. Total cardiopulmonary bypass and aortic cross-clamp times and return to bypass were recorded. Reoperations performed during the 6-month follow-up were noted.

Echocardiograms

Echocardiographic data were collected as previously reported [4, 5]. Local investigators reviewed all preoperative echocardiograms. Echocardiograms performed 1 month postoperatively were reviewed locally for the participants retrospectively enrolled and in the core laboratory for those prospectively enrolled. Only prospectively enrolled patients had 6-month echocardiograms and all were reviewed in the core laboratory. The degree of LAVVR was graded as none/trace, mild, moderate, or severe (moderate or severe was considered significant). Color Doppler imaging was also used to demonstrate a residual ASD or VSD (diameters > 3 mm were considered significant). Pulsed Doppler interrogation was used to evaluate LAVV inflow and left ventricular (LV) outflow. A LAVV mean inflow gradient of 7.5 mm Hg or higher and a LV outflow peak gradient of 20 mm Hg or higher were considered significant.

Statistical Analysis

Descriptive statistics are presented as mean $\pm$ standard deviation, median and interquartile range, or frequencies and percentages. The distributions of categoric variables by AVSD subtype were compared using the Fisher exact or χ^2 test. Ordinal variables were analyzed using the Mantel-Haenszel test for linear trend. Analysis of variance and the Kruskal-Wallis test were used to compare normally distributed and skewed variables by subtype, respectively. The one-sample t test was used to determine whether change in the weight-for-age z score differed from 0 for each subtype. Univariate and multivariable logistic regressions were used to identify patient and surgical factors associated with the outcome of moderate or severe LAVVR at the 6-month follow-up. With all participants combined, interactions between defect subtype and prespecified factors (age at operation, annuloplasty, and preoperative and 6-month postoperative LAVVR) were evaluated. Linear regression was used to identify predictors of log-transformed hospital days in patients who underwent operations age 1 month or older.

All variables significant at the 0.20 level in univariable analysis were evaluated for inclusion in multivariable regression models. A test of interaction was used to assess whether the association of age at repair with hospital outcomes depended on the number of associated cardiac anomalies. Exploratory data analyses were conducted using generalized additive models. Stepwise multivariable linear regressions were performed for each defect subtype to explore factors associated with the 6-month change in weight-for-age z score. The R^2 values reported are adjusted R^2 and maximum rescaled R^2 for linear and logistic regression, respectively. All analyses were performed using SAS 9 software (SAS Institute, Inc, Cary, NC).

Results

Participants

The study enrolled 215 patients (Table 1) with the following AVSD subtypes: PAVSD, 60 (28%); TAVSD, 27 (13%); CAVSD, 120 (56%), and canal-type VSD, 8 (4%). Compared with other subtypes, PAVSD participants were significantly older at the time of repair. The CAVSD and canal-type VSD groups had a higher percentage of children with trisomy 21, significantly lower average weight-

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Table 1. Patient Characteristics and Hospitalization Statistics by Atrioventricular Septal Defect Subtype

Variable	PAVSD (n = 60)	TAVSD (n = 27)	CAVSD (n = 120)	Canal-Type VSD (n = 8)	*p* Value
Age at repair, years					<0.001
Mean ± SD	3.6 ± 3.3	1.4 ± 1.30	0.3 ± 0.2	0.4 ± 0.1	
Median (IQR)	3.1 (1.3, 4.7)	0.7 (0.5, 2.5)	0.3 (0.2, 0.4)	0.4 (0.3, 0.4)	
Male sex, %	55	41	41	38	0.31
Ethnicity/race, %					0.30
Hispanic	10	11	13	0	
Non-Hispanic white	78	63	71	63	
Non-Hispanic black	7	11	13	25	
Non-Hispanic other	5	15	3	13	
Trisomy 21, %	28	44	80	75	<0.001
Cleft closure, %	98	100	93	75	0.04
Annuloplasty, No. (%)	16 (27)	6 (22)	29 (24)	1 (13)	0.91
Bypass time, min	73 ± 26	95 ± 41	112 ± 36	120 ± 22	<0.001
Return bypass, %	2	7	6	13	0.23
Cross-clamp time, min	47 ± 18	64 ± 29	82 ± 31	81 ± 20	<0.001
Circulatory arrest, %	3	0	8	13	0.24
Additional operations, %	12	26	48	50	<0.001
Total ventilator days					<0.001
Mean ± SD	2.9 ± 10.3	2.0 ± 1.6	6.0 ± 14.9	3.6 ± 2.9	
Median (IQR)	1 (1, 1)	1 (1, 3)	2 (1.5, 4)	2 (2, 4.5)	
Total LOS, days					
Intensive care unit					<0.001
Mean ± SD	4.5 ± 11.5	3.1 ± 2.0	8.6 ± 16.8	5.8 ± 3.7	
Median (IQR)	2 (2, 3)	2 (2, 4)	4 (3, 6)	4.5 (3, 8)	
Hospital					
Mean ± SD	7.0 ± 12.0	8.1 ± 6.1	14.5 ± 20.4	8.6 ± 3.7	
Median (IQR)	5 (4, 5)	5 (5, 11)	8 (6, 12)	7.5 (5.5, 11.5)	<0.001

CAVSD = complete atrioventricular septal defect; IQR = interquartile range; LOS = length of stay; PAVSD = partial atrioventricular septal defect; SD = standard deviation; TAVSD = transitional atrioventricular septal defect; VSD = ventricular septal defect.

for-age *z* scores at operation, and significantly more participants underwent repair of additional cardiac lesions. Subgroups differed in mean cardiopulmonary bypass and cross-clamp times ($p < 0.001$).

Surgical approaches used in the CAVSD group included double-patch (72%), single-patch (18%), and the Australian technique (10%), and their effects on outcomes have been reported [5]. When pairwise comparisons of the four group means were performed, PAVSD participants had the lowest bypass and cross-clamp times (for both: $p = 0.007$ vs TAVSD; $p < 0.001$ vs CAVSD and vs canal-type VSD). Mean times were lower in TAVSD participants ($p = 0.02$) than in CAVSD participants ($p = 0.002$), but were similar in CAVSD ($p = 0.49$) and canal-type VSD ($p = 0.96$). Cleft closure was performed in 75% with a canal-type VSD and in 93% or more of all other subtypes.

Postoperative Course

Six patients died, 1 with PAVSD and 5 with CAVSD, all with associated pathology [4, 5]. Median ventilation, intensive care unit, and hospital days were significantly longer in the CAVSD group compared with PAVSD and TAVSD groups (Table 1). At the 6-month follow-up, 1 TAVSD and 1 CAVSD patient had a residual ASD (> 3 mm) and 1 CAVSD patient had a residual VSD (> 3 mm). At the 6-month follow-up, 1 PAVSD patient had subaortic stenosis (peak gradient, 39 mm Hg), 1 TAVSD and 3 CAVSD patients had LAVV stenosis (mean gradient range, 7.7 to 10.9 mm Hg), and 1 CAVSD patient required a pacemaker for complete heart block.

Perioperative complications have been reported previously [4, 5]. Sepsis and pleural effusions were the most frequent complications for PAVSD (7%) and TAVSD (5%). For the CAVSD group, gastrointestinal complications (15%) and pleural effusions (14%) were the most common. Pleural effusion (2 of 8) was the most frequently reported complication for canal-type VSD.

Left AV Valve Regurgitation

Preoperatively, TAVSD subjects had the highest prevalence (44%) of moderate or severe LAVVR (Table 2) and canal-type VSD participants had the lowest (0 of 8). Reoperation for residual LAVVR within 6 months of initial repair was performed in 7 patients (Table 3). The 1-month postoperative echocardiogram revealed a decrease in the prevalence of moderate or severe LAVVR for PAVSD and TAVSD subtypes but an increase in

Table 2. *Patients With Moderate of Severe Left Atrioventricular Valve Regurgitation by Atrioventricular Septal Defect Subtype*

Timing of Echocardiogram[a]	PAVSD	TAVSD	CAVSD	Canal-Type VSD	*p*-Value
Preoperative					
Total No.	60	27	120	8	
Moderate/severe LAVVR	19 (32)	12 (44)	25 (21)	0 (0)	0.02
1-month postoperative					
Total No.	60	27	119	8	
Moderate/severe LAVVR	11 (18)	6 (22)	29 (24)	2 (25)	0.83
6-months postoperative					
Total No.	38	21	83	7	
Moderate/severe LAVVR	11 (29)	7 (33)	18 (22)	2 (29)	0.61

[a] Data are presented shown as number (%).

CAVSD = complete atrioventricular septal defect; LAVVR = left atrioventricular valve regurgitation; PAVSD = partial atrioventricular septal defect; TAVSD = atrioventricular septal defect; VSD = ventricular septal defect.

CAVSD and canal-type VSD subtypes. At the 6-month follow-up, 39 of 150 (26%) overall had moderate or severe LAVVR. The prevalence of LAVVR was similar among centers. CAVSD Rastelli classifications (A = 63; B = 2; C = 27; unclassified = 28) were not associated with the prevalence of moderate or severe LAVVR at the 1-month or 6-month follow-up.

The rate of annuloplasty use was similar among AVSD subtypes ($p = 0.91$; Table 1), but annuloplasty use varied widely, from 3% to 49%, among the 7 centers ($p < 0.001$). Patients with moderate or severe LAVVR preoperatively were more likely to undergo annuloplasty than those with mild or none LAVVR (38% vs 20%; Table 4), with an odds ratio of 2.5 for annuloplasty (95% confidence interval [CI], 1.27 to 4.83, $p = 0.008$). Despite this, the use of annuloplasty was not associated with a significant reduction in the prevalence of moderate or severe LAVVR 6 months postoperatively (24% in the annuloplasty group vs 26% in the no annuloplasty group, $p = 0.83$).

Multivariable analysis showed that the strongest predictors of LAVVR at the 6-month follow-up varied among subtypes (Table 5). For PAVSDs, moderate or severe LAVVR on the preoperative and 1-month postoperative echocardiograms independently predicted moderate or severe LAVVR at 6 months. Older age at repair was the only independent predictor of significant LAVVR for TAVSD, and LAVVR at the 1-month postoperative echocardiography was the only independent predictor of LAVVR for CAVSD at the 6-month follow-up. To maximize the power of LAVVR predictors across all subtypes, modeling using all participants was conducted, with interaction terms evaluated for subtype by age at repair, preoperative LAVVR, 1-month postoperative LAVVR, and weight-for-age z score at operation. No interactions were significant.

The multivariable model based on all patients identified two independent predictors associated with moderate or severe LAVVR at the 6-month follow-up for all subtypes: older log(age) at repair ($p = 0.02$) and moderate or severe LAVVR on the 1-month postoperative echocardiography study ($p < 0.001$). Annuloplasty, center, return to bypass, degree of preoperative LAVVR, and subtype were not independent predictors of moderate or severe LAVVR at 6 months.

The interaction between age at repair and subtype was in part not significant because most patients (CAVSD) had repairs only in infancy. Therefore, we conducted nonparametric regression analyses of age at repair vs 6-month LAVVR separately for each subtype to qualitatively depict associations with age (Fig 1). The relationship between the highest prevalence of LAVVR at 6 months postoperatively and age at repair varied by

Table 3. *Description of Reoperations for Left Atrioventricular Valve Regurgitation*

	Moderate/Severe LAVVR			
AVSD Subtype	Pre-op	1-mon post-op	Annuloplasty	Reoperation
CAVSD	Yes	Yes	Yes	LAVV replacement
CAVSD	No	Yes	No	Repeat LAV valvuloplasty, LAVV replacement × 2
CAVSD	No	Yes	No	Repeat LAV valvuloplasty
CAVSD	Yes	Yes	Yes	Repeat LAV valvuloplasty
PAVSD	No	Yes	No	Repeat LAV valvuloplasty
TAVSD	Yes	Yes	Yes	Repeat LAV valvuloplasty
TAVSD	No	Yes	No	Repeat LAV valvuloplasty

AVSD = atrioventricular septal defect; CAVSD = complete atrioventricular septal defect; LAV =left atrioventricular; LAVV = left atrioventricular valve; LAVVR = left atrioventricular valve regurgitation; PAVSD =partial atrioventricular septal defect; TAVSD = transitional atrioventricular septal defect.

Table 4. Patients With Annuloplasty by Subtype and Left Atrioventricular Valve Regurgitation Grade

Group	PAVSD No. (%)	TAVSD No. (%)	CAVSD No. (%)	Canal-type VSD No. (%)	Overall No. (%)
All patients	16/60 (27)	6/27 (22)	29/120 (24)	1/8 (13%)	52/215 (24)
Pre-op LAVVR status					
Patients with moderate/severe	8/19 (42)	5/12 (42)	8/25 (32)	0/0 (—)	21/56 (38)
Patients with mild/none	8/41 (20)	1/15 (7)	21/95 (22)	1/8 (13)	31/159 (20)
p Value	0.11	0.06	0.30	NA	0.01

CAVSD = complete atrioventricular septal defect; LAVVR = left atrioventricular valve regurgitation; PAVSD = partial atrioventricular septal defect; TAVSD = transitional atrioventricular septal defect; VSD = ventricular septal defect.

subtype (Fig 1). For PAVSD, the peak prevalence clearly occurred in those undergoing repair after 4 years of age. For TAVSD and CAVSD, however, the prevalence of LAVVR increased linearly with increasing age at repair. These two subgroups accounted for 104 of 142 patients with 6-month LAVVR data and explain why model 4 (Table 5) found log(age) was significantly associated with outcome in logistic regression using the combined cohort.

Postoperative Weight Gain

All subtypes showed a similar increase in weight-for-age z scores after repair, with a mean change of 0.5 to 1.3 ($p = 0.17$, Fig 2), and the overall 95% CI for the improvement in weight-for-age z score was 0.94 to 1.17. Separate stepwise multivariable linear regression analyses were performed by subtype (Table 6) to explore potential factors associated with the change in weight-for-age z score (z score at 6 months minus z score at operation). Stratified analyses showed factors associated with weight gain varied among subtypes. For PAVSD, weight improved the most in those who had longer intensive care stays ($p = 0.004$). For TAVSD, the presence of trisomy 21 and no annuloplasty at repair were independently associated with a greater improvement in weight-for-age z score ($p < 0.001$ for both) and, along with center adjustment, accounted for 96% of the variation in outcome. For CAVSD, shorter hospital stay ($p = 0.02$) and lower preoperative weight-for-age z score ($p < 0.001$) were independently associated with greater postoperative weight gain, accounting for 39% of the variation in outcome.

Resource Utilization

Median days of ventilation, intensive care, and hospitalization varied by subtype ($p < 0.001$, Table 1). Predictors of hospitalization days were examined after excluding 5 patients who required urgent repair because of associated defects before they were 1 month old (Table 7). No significant interaction between subtype and potential predictors was found. Analysis using data from all subtypes showed no cleft closure, longer bypass times, moderate or severe LAVVR on 1-month postoperative echocardiogram, and younger log(age) at repair were independently associated with longer hospitalization ($R^2 = 0.31$). Annuloplasty was associated with more hospital days only for CAVSD (17.9 ± 24.9 vs 11.8 ± 16.5, $p = 0.003$).

Comment

Atrioventricular septal defects are one of the congenital heart defects for which outcomes are used to assess the pediatric heart surgeon's skill as well as institutional expertise [3–5]. This multicenter observational study reports contemporary outcomes for all AVSD subtypes and analyzes patient characteristics, resource utilization, and practice variation as potential predictors of these outcomes.

After adjusting for subtype, centers were similar in age at repair and days of ventilation, intensive care, and hospitalization. The outcomes at 6 months compared favorably with those reported from single-center studies. Specifically, overall mortality was low, with only 6 deaths (3%) compared with the 7% to 15% mortality rate reported from other centers [9–13]. Septal defects (> 3 mm) occurred in 2% at the 6-month follow-up, similar to previous reports where residual defects occurred in 5%

Table 5. Predictors of Left Atrioventricular Valve Regurgitation 6 Months After Atrioventricular Septal Defect Repair

Logistic Regression Model	No.	OR (95% CI)	R^2	*p* Value
Model 1: Partial AVSD	38		0.41	
Moderate/severe LAVVR pre-op		19.2 (1.9–188)		0.01
Moderate/severe LAVVR 1-mon post-op		19.6 (1.6–241)		0.02
Model 2: Transitional AVSD	21		0.25	
Log(age)-at-repair		3.8 (0.9–15)		0.06
Model 3: Complete AVSD	83		0.19	
Moderate/severe LAVVR 1-mon post-op		6.9 (2.1–22)		0.001
Model 4: For all subtypes	149		0.17	
Log(age)-at-repair		1.6 (1.1–2.2)		0.02
Moderate/severe LAVVR 1-mon post-op		4.8 (2.1–11		<0.001

AVSD = atrioventricular septal defect; CI = confidence interval; LAVVR = left atrioventricular valve regurgitation; OR = odds ratio.

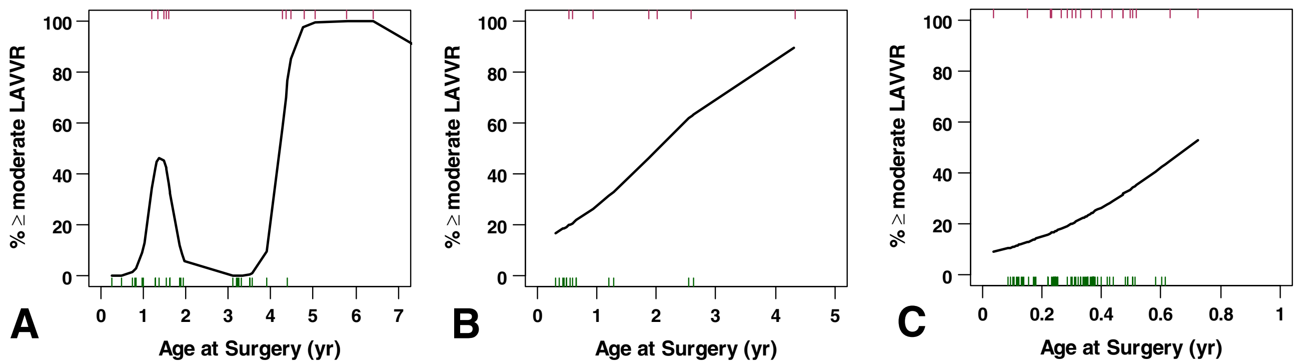

Fig 1. Smoothed nonparametric regression estimates depict the relationship between age-at-repair (years) and the percentage of subjects with moderate/severe (≥ moderate) left atrioventricular valve regurgitation (LAVVR) for the (A) partial, (B) transitional, and (C) complete subtype. Canal-type VSD was not included because of the small sample size. The green hash marks denote age at repair for patients with moderate/none LAVVR. The purple hash marks denote age at repair for patients with moderate/severe LAVVR.

or fewer [12, 13]. Subaortic stenosis and LAVV stenosis were uncommon and lower than the 5% previously reported for these lesions [14], but this low prevalence may be attributed to the short duration of follow-up.

Residual LAVVR remains the Achilles' heel of AVSD repair, with 3% (7 of 215) requiring reoperation and 26% with moderate or severe LAVVR by the 6-month follow-up. AVSD subtype and center were not independent predictors of the prevalence of moderate or severe LAVVR at 6 months. Despite the nearly uniform adoption of complete LAVV cleft closure, the prevalence of significant LAVVR has remained unchanged during the past decade [9, 15, 16]. To decrease LAVVR, some surgeons have advocated the addition of an annuloplasty [16, 17], but this remains controversial and its use varied widely among the 7 centers. For all subtypes, some surgeons performed annuloplasty prophylactically, even in those with mild or no preoperative LAVVR, some used it only if the preoperative LAVVR was graded as moderate or severe, and others did not use it at all. Regardless of the reason for its use, however, we were unable to demonstrate that annuloplasty decreased the prevalence of moderate or severe LAVVR at 6 months postoperatively. In fact, for reasons that are unclear, annuloplasty appeared to have some negative aspects in this study, where its use was associated with poorer weight gain in TAVSD patients and increased resource utilization in the CAVSD group.

Older log(age) at repair and the presence of moderate or severe LAVVR within 1 month of the operation predicted moderate or severe LAVVR at the 6-month follow-

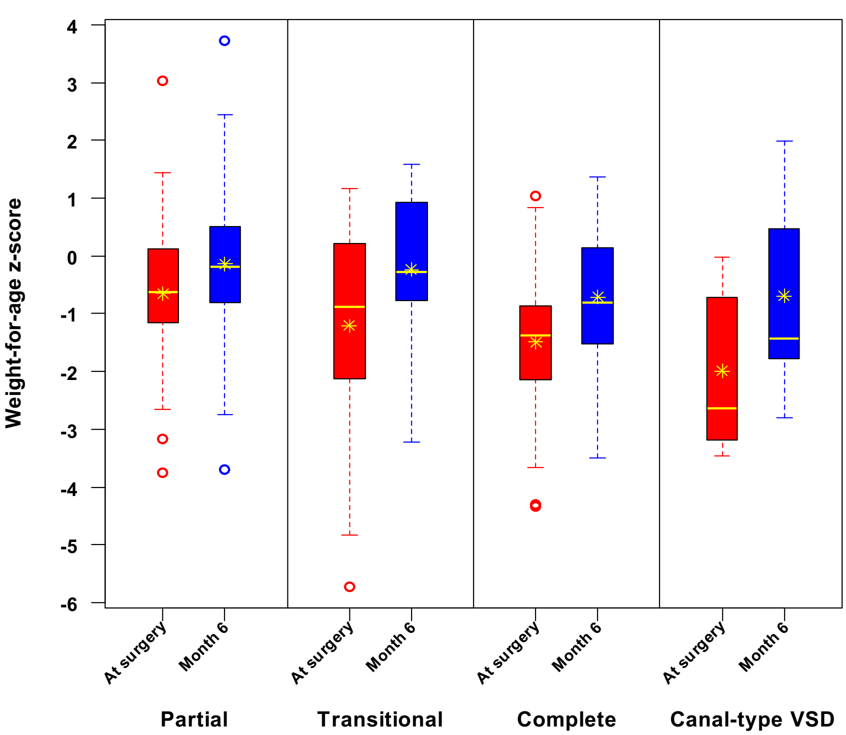

Fig 2. Weight-for-age z score by atrioventricular septal defect (AVSD) subtype and time point (paired data). The horizontal line in the middle of each box indicates the 50th percentile; the top and bottom borders of the box mark the 75th and 25th percentiles, respectively; and the whiskers mark the 90th and 10th percentiles. The asterisk (*) denotes the mean. Red denotes the z score at operation, and blue denotes the z score at 6 months. Growth improved significantly in patients with all subtypes.

up. The prevalence of moderate or severe LAVV regurgitation clearly peaked after later operations for PAVSD but increased linearly for TAVSD/CAVSD. Other investigators have noted this trend and advocated earlier repair [9, 10]. Despite earlier repair during the past decade, however, the prevalence of postoperative LAVVR has remained unchanged [9, 15, 16]. Our data do not permit us to determine the ideal time of repair to minimize postoperative LAVVR.

Patient characteristics differed by subtype. CAVSD and canal-type VSD had a significantly more patients with trisomy 21, more associated cardiac defects, and longer cardiopulmonary bypass and cross-clamp times compared with PAVSD or TAVSD. We confirmed the commonly held belief that CAVSDs were more likely to have lower weight-for-age *z* scores at the time of repair. More surprising is the finding that weight gain significantly improved 6 months after repair in all subtypes, including those with PAVSD, and that all subtypes showed a similar increase in weight-for-age *z* scores. Although the predictors of improvement in weight varied with subtype, all subtypes likely share the common characteristic of being more nutritionally depleted before the operation and thus are able to display better catch-up growth after repair.

This study has several limitations: First, the small numbers of each subtype limited the ability to detect some associations and interactions of outcome predictors and subtype.

Second, because our data were collected both prospectively and retrospectively, all measurements could not be centrally interpreted, and 6-month postoperative echocardiographic data were not available for some participants. In addition, reliable and validated echocardiographic methods for quantitative evaluation of LAVVR grade are not available for children, particularly in the setting of multiple or eccentric jets characteristic of the repaired AVSD valve. We used qualitative assessment of the color Doppler jet because this was the standard for clinical decision making in all 7 centers. This has also been the most commonly used method for grading LAVVR in other AVSD studies, allowing for comparisons to be made.

Finally, we were unable to explore the effect of variations in annuloplasty technique on valve function because annuloplasty details were not uniformly available from the operative reports.

In conclusion, survival in the current era is excellent, with few residual defects after surgical repair for all AVSD subtypes at a similar age of repair among the 7 centers. After accounting for age at repair, we found subtype and center were not important prognostic indicators of resource utilization, postoperative LAVVR, or improvement in growth. Significant postoperative LAVVR is the most common sequela, with a similar prevalence across the centers at 6 months. Annuloplasty use varied among centers and failed to decrease the prevalence of moderate or severe LAVVR at the 6-month follow-up in this cohort. Older age at repair was an independent predictor of moderate or severe LAVVR. Further investigation is needed to determine if intervening in center practice variation or earlier referral of subtypes traditionally repaired at an older age can affect the relatively high prevalence of postoperative LAVVR.

Table 6. Change in Weight-for Age Z Scores for Patients With Paired Data at 6 Months

Model	Estimate	Adjusted Mean	R^2	*p* Value
Model 1: Partial AVSD			0.18	
Log(intensive care days)	0.54			0.004
Model 2: Transitional AVSD			0.96	
Site				<0.001
Trisomy 21		1.22		<0.001
Annuloplasty		−1.49		<0.001
Model 3: Complete AVSD			0.39	
Log(hospital days)	−0.42			0.02
Weight-for-age *z* score at operation	−0.56			<0.001
Model 4: All patients (N =152)			0.28	
Weight-for-age *z* score at operation	−0.40			<0.001

AVSD = atrioventricular septal defect.

Table 7. Multivariable Model for Log (Hospital Length of Stay) After Atrioventricular Septal Defect Repair (N = 210)

Model (R^2 = 0.31)	Variable Estimate	Adjusted Mean	*p* Value
Cleft closure			
Yes	−0.53	2.06	<0.001
No		2.59	
Bypass time, min	0.005		<0.001
LAVVR on 1-month post-op echo	0.21		0.01
Moderate/severe		2.42	
None/mild		2.22	
Log(age-at-repair), days	−0.20		<0.001

LAVVR = left atrioventricular valve regurgitation.

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DISCUSSION

DR DUKE CAMERON (Baltimore, MD): You may have mentioned this, were all clefts closed in all of these cases?

DR KAZA: Ninety-three percent of the patients in this study had their cleft closed, Dr Cameron. This was not closed in the rare patient with single papillary muscle or deficient left lateral leaflet.

DR HAROLD BURKHART (Rochester, MN): Very nice report. Do you have any information on the type of repair utilized for the complete atrioventricular (AV) canal defects; 1 patch vs 2, and if the bridging leaflet was divided?

DR KAZA: Harold, that is actually going to come out in print where the Pediatric Heart Network did a detailed analysis of the complete atrioventricular septal defect (AVSD) group looking at the surgical technique. The specific type of surgical repair for complete atrioventricular septal defect (CAVSD) is beyond the scope of this project, but it should be coming out in the *Journal of Thoracic and Cardiovascular Surgery* in the next couple of months.

DR ROBERT JAQUISS (Durham, NC): This was very interesting to me. I had expected to see that preoperative AV valve regurgitation was predictive. And I was also a little surprised that being older is predictive of more AV regurgitation afterwards, because I would have predicted the kids that come to surgery earlier, especially the complete canals where the doctors that are referring are always wondering about when to refer them, are more likely to refer somebody who has got worse LV valve regurgitation. And I was also gratified to see that I am not the only one that has a cohort of patients wandering out there at 6 months with significant mitral regurgitation. It's just a comment.

DR KAZA: Dr Jaquiss, I think that is a very important point. I think that certain subtypes of AVSD patients do benefit from early referral. The one thing I have noticed personally is that the older transitional canals that come for repair, where they are referred for repair at 5 to 6 years, is that the leading edges of the bridging leaflets have contracted and rolled in and it is difficult to bring them together with a reliable degree of coaptation. And it almost feels like you are distorting the annulus. Those are the patients that have residual regurgitation as well.

DR CAMERON: I thought it was interesting that none of your inlet VSDs had mitral regurgitation before repair. And was it a quarter of them had moderate to severe after repair?

DR KAZA: The study included a total of 8 patients with inlet VSDs.

DR CAMERON: Is that the same phenomenon we were talking about before where closing septal defects caused mitral regurgitation, or is it just a cleft closure that's gone amiss?

DR KAZA: I think that the high percentage of residual regurgitation in these patients is related to the power. There were 8 patients with inlet VSDs and 2 of them did develop AV valve insufficiency postoperatively, and that comes up to be a huge percentage. I am not sure about the precise mechanism as to why they developed the residual insufficiency.