

Early Repair of Complete Atrioventricular Septal Defect is Safe and Effective

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Background. Surgical repair of complete atrioventricular septal defect (CAVSD) is a well-established procedure performed on young children. Our hypothesis is that with modern techniques, the current risks of CAVSD repair in children aged younger than 3 months and in children older than 3 months are equal.

Methods. This was a retrospective review of 65 infants and children with a mean age of 10.9 months (range, 1 month to 15.5 years) who underwent CAVSD repair from 1990 to 2004. Twenty-six repairs (40%) were done on or before 3 months of age (group A) and 39 repairs (60%) were done after 3 months of age (group B). In all patients, the ventricular septal defect was repaired with an individualized approach according to each patient's specific anatomy: direct suturing without a patch, interposition of a small pericardial patch with a running suture, or both. The atrioventricular commissure was closed with inter-

rupted sutures, and all atrial defects were closed with a pericardial patch. Data were analyzed using the χ^2 analysis and the Fisher exact test.

Results. Three hospital deaths occurred (<30 days), 2 in group A and 1 in group B (7.7% vs 2.6%, respectively, $p = 0.33$). One death in group A occurred during another noncardiac surgery. Early reoperation (<1 year of initial surgery) for residual ventricular septal defect or significant mitral regurgitation, or both, occurred in 3 group A patients and in 4 group B patients (11.5% versus 10.3% respectively, $p = 0.68$).

Conclusions. These results suggest that repair of CAVSD defects in children 3 months of age or younger had similar outcomes compared with those who underwent surgical repair after 3 months of age.

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Complete atrioventricular septal defect (CAVSD) is a term used to describe a constellation of congenital cardiac structural abnormalities characterized by an ostium primum atrial septal defect, a common atrioventricular (AV) valve, and a nonrestrictive deficiency of the ventricular septal inflow. The latter, along with the amount of AV valve regurgitation, determines the onset of symptoms. Patients with mild AV valve regurgitation and high pulmonary vascular resistance can remain relatively asymptomatic until they start developing cyanosis from advanced pulmonary vascular disease. In contrast to that, if the peripheral vascular resistance falls, as it normally does at 6 weeks of life, large left-to-right shunts develop through the septal defects. This in turn leads to signs and symptoms of congestive heart failure, which can also develop in the setting of severe AV valve regurgitation.

About half of these patients, if left untreated, will die within the first year of life, usually from heart failure or respiratory tract infections [1]. Pulmonary hypertension invariably develops in those who survive [2]. The pulmonary vascular disease becomes irreversible over time, thus precluding surgical treatment; hence, it stands to

reason that CAVSD should be repaired before the onset of irreversible pulmonary hypertension [3, 4]. This is preferably scheduled before the sixth to twelfth months of life.

With improvements in anesthetic and intensive care, surgical repair of CAVSD is now the treatment of choice and is currently performed in infancy [3–8]. In a subset of infants, however, early congestive heart failure develops within the first few weeks of life that is usually not controlled with medical therapy alone. It is in these patients that controversy exists about the ideal timing of surgery [7, 9]. Surgical correction in these infants involves the handling of smaller and more delicate cardiac structures, but delaying it may lead to devastating outcomes.

Unfortunately, it is not just timing of surgery that is controversial in these fragile infants. Many surgical techniques have been proposed for the repair of these defects [10], but a more customized approach may be more appropriate given their highly variable morphology [11–13]. We used a simpler technique that could be customized to each patient's specific anatomic arrangement. Thus, the purpose of this study was to test the hypothesis that in the modern era, using the techniques described in this study, early (<3 months old) repair of CAVSD is safe and effective.

Patients and Methods

Approval to collect and analyze data from a retrospective chart review was received from the University of Virginia

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Table 1. Preoperative Data

	Group A ^a (n = 26) (%)	Group B ^b (n = 39) (%)	p Value
Gender			
Males	10 (38)	17 (44)	0.344
Females	16 (62)	22 (56)	
Down syndrome	18 (69)	36 (92)	0.001
≥Moderate LAVVR	4 (15)	10 (26)	0.157

^a Early (≤ 3 months old) complete atrioventricular septal defect repair.

^b Late (>3 months old) complete atrioventricular septal defect repair.

LAVVR = left atrioventricular value regurgitation.

Human Investigation Committee/Institutional Review Board, which waived the need for obtaining patient consent for this study. We reviewed the records of infants and children who underwent CAVSD repair at the University of Virginia Health System between 1990 and 2004. All patients in whom an individualized approach was used to perform the procedure, as described previously [14], were included in the study. This technique was first initiated in 1990 and so that was chosen as the starting date of the study. The study excluded patients with associated complex cardiac anomalies (ie, transposition of the great arteries, tetralogy of Fallot) because they would not have comparable anatomy or physiology, resulting in an altered natural history and surgical management of CAVSD.

Table 1 presents the preoperative data for early (≤ 3 months old, group A) and late (>3 months old, group B) CAVSD repair.

Surgical Procedure

Patients were placed on standard cardiopulmonary bypass with two venous cannulae. Moderate hypothermia was induced, and antegrade cold blood cardioplegia was administered after cross-clamp placement. The first step of the repair was to close the left AV commissure with interrupted 5-0 polypropylene sutures through a right atriotomy. After ensuring coaptation, the ventricular septal defect was assessed.

An individualized approach was used in all patients to repair the ventricular septal defect according to each patient's specific anatomy: direct suturing without a patch infrequently, or interposition of a pericardial patch with a running suture. Direct suturing was done with 5-0 polypropylene sutures in an interrupted horizontal mattress fashion if this septal defect could be closed without tension. The sutures were placed through the right ventricular side of the ventricular septal crest and then through the common valve leaflets. If that would result in undesirable tension, distortion of the AV valves, or if the defect was large, a bovine pericardial patch was sutured in between the common AV valve leaflets and the ventricular septal crest. This was done with a running polypropylene suture. Not infrequently, an amalgamation of these two methods could be used, with direct suturing at the edges of the defect leaving room for a

smaller patch in the larger, central area of the ventricular septal defect.

Care was taken in all of these techniques to ensure that most of the leaflet tissue was made a part of the newly constructed left AV valve. This was done by taking up more tissue from the right half of the common leaflets. Nevertheless, one has to be careful doing this when the right-sided section of the AV valve is small because it may result in a tricuspid orifice that is too small.

The atrial component was then closed with a second patch. However, before that was done, the ventricular outflow tract patency was established by passing a probe into it through the left AV valve orifice. All atrial defects were closed with a bovine pericardial patch using a running 5-0 polypropylene suture. This was done preferentially having the coronary sinus draining into the left atrium so as to steer clear of the conduction tissue. However, the coronary sinus was directed into the right atrium when there was a persistent left superior vena cava present. Finally the right atriotomy was closed using a running 5-0 polypropylene suture. The repair was assessed intraoperatively with transesophageal echocardiography.

Statistical Analysis

A binary logistic regression analysis was performed for both mortality and reoperation separately. In addition, univariate statistics using either a χ^2 analysis or a Fisher exact test were performed comparing the variables age, reoperation, mortality, Down syndrome, gender, and the degree of postoperative mitral regurgitation. A significant difference was indicated at $p \leq 0.05$. All analyses were performed using the statistical software SPSS (SPSS Inc, Chicago, IL). The binary logistic regression failed to show any significant predictors for the variables mortality and reoperation.

Results

The repair was performed on 65 infants and children, with a mean age of 10.9 months (range, 1 month to 15.5 years). Twenty-six repairs (40%) were done on or before 3 months of age (group A) and 39 repairs (60%) were done after 3 months of age (group B). The mean ages of patients were 2.14 months in group A (range, 1 to 3 months) and 16.76 months in group B (range, 3.5 to 186 months). The yearly distribution of patients stratified by age group is shown in Figure 1.

All of the patients were studied with preoperative echocardiography. Additional evaluation with preoperative cardiac catheterization was performed in 13 cases. Cardiovascular anomalies associated with CAVSD included patent ductus arteriosus (19 patients), dextrocardia (1 patient), situs inversus (1 patient), and persistent left superior vena cava with drainage into the coronary sinus (5 patients).

The decision to operate on these children was made on their first presentation with symptoms of heart failure, which included failure to thrive, weight loss, and recurrent respiratory tract infections. No patient in this series

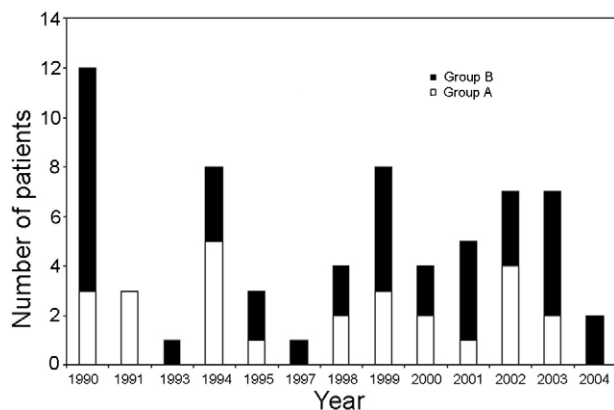


Fig 1. Yearly distribution of patients stratified by age group. Group A = clear bars; group b = solid bars.

had previously undergone palliative pulmonary artery banding, and all repairs were done electively. Similar numbers of patients in both groups had moderate or severe left AV valve regurgitation. There were no differences in gender between the two groups, and group A had fewer patients with Down syndrome than group B (69% versus 92%, $p = 0.001$).

The postoperative data are summarized in Table 2. A similar number of ventricular septal defects were closed without a patch in both groups (13.3% in group A and 12.5% in group B). Overall hospital (<30 days) mortality was 4.6%. Two (7.7%) of 26 patients died in group A and 1 (2.6%) of 39 patients in group B ($p = 0.33$). One of the deaths in group A occurred in an infant due to complications of a Nissen fundoplication. The cardiac repair was intact in both of the group A patients who died, and there was no evidence of a residual shunt or left ventricular outflow tract obstruction. The patient who died in group B did, however, have moderate outflow tract obstruction and mitral valve regurgitation.

Early reoperation (≤ 1 year of initial surgery) for residual ventricular septal defect or significant mitral regurgitation, or both, occurred in 3 patients in group A and 4 patients in group B (11.5% versus 10.3%, respectively, $p = 0.68$). This brought the overall reoperation rate to 10.8%. All of these patients recovered and were alive at the time of follow-up.

Table 2. Postoperative Data

	Group A ^a (n = 26) (%)	Group B ^b (n = 39) (%)	p Value
Hospital mortality	2 (7.7)	1 (2.6)	0.33
Early reoperation	3 (11.5)	4 (10.3)	0.68
\leq Mild LAVVR	19 (73)	29 (74)	
Moderate LAVVR	6 (23)	9 (23)	0.76
Severe LAVVR	1 (4)	1 (3)	

^a Early (≤ 3 months old) complete atrioventricular septal defect repair.

^b Late (>3 months old) complete atrioventricular septal defect repair.

LAVVR = left atrioventricular valve regurgitation.

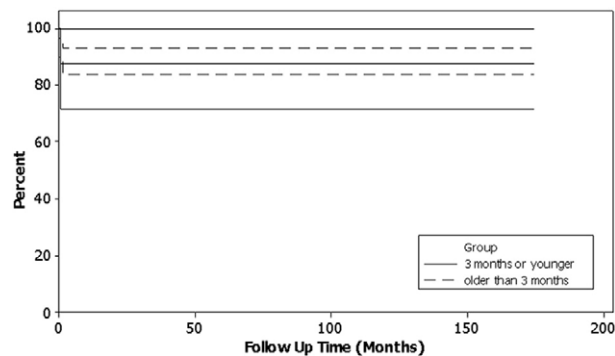


Fig 2. Survival plot for follow-up time (months) determined by Kaplan-Meier method, with 95% confidence intervals shown by upper and lower lines. (Solid line = patients 3 months or younger; dashed line = patients older than 3 months.)

One further reoperation was required in group A 2 years after the initial surgery for residual severe mitral regurgitation with closure of a secundum atrial septal defect. This was also the length of time after the initial surgery that a patient in group B had a reoperation for residual ventricular septal defect.

Three complications occurred in group A, all within 1 month of the initial surgery, which required intervention: heart block requiring a pacemaker, percutaneous drainage of a pericardial effusion, and a pleural effusion. Similarly, four complications, all due to heart block, occurred in group B that required the insertion of a pacemaker. These interventions took place in 2 patients at 1 month after the initial operation, in another patient at 8 months, and at 2.5 years after repair of the CAVSD in the fourth patient.

All patients were followed-up with periodic clinical examination and echocardiography, and 100% of patients had at least short-term follow-up. A Kaplan-Meier survival plot for follow-up time is shown in Figure 2. The median follow-up time for these patients was 48 months (range, 0.5 to 174.5 months). The median follow-up time was 44 months for group A (range, 0.6 to 150.7 months) and 48 months for group B (range, 0.5 to 174.5 months). Pneumonia caused one late death in group B.

Echocardiography examination at the latest clinic follow up revealed absent-to-mild residual left AV valve incompetence in 19 patients in group A (73%) and 29 in group B (74%; $p = 0.76$). Hence, no significant differences in operative mortality, early reoperation, or residual left AV valve regurgitation were observed between the two groups studied. It is conceivable that a difference may not have surfaced owing to the low power of this study.

Comment

CAVSDs are associated with high-flow systemic pressures in the pulmonary vasculature leading to fibrosis and intimal hyperplasia. This eventually leads to a reduction in the total cross-sectional area of the pulmonary vascular bed. Surgery within 6 months usually prevents these irreversible obliterative changes [15, 16], and there-

fore, this pathology should be repaired as early as possible. In addition, pulmonary hypertension can occur by the third month of life so we chose this age to be the division point of the two groups in our study.

Palliation with pulmonary artery banding is now seldom indicated and has now been abandoned for a single-stage definitive surgical repair [10]. In recent years, there has been a better understanding of the surgical anatomy of CAVSD as well as improvements in surgical techniques and postoperative care. This in turn has led to a decrease in the mortality rate among this patient population [3, 9]. Several previous studies demonstrated that repair at an early age was beneficial [9, 17]. In fact, early correction could partially eliminate the incidence of left AV valve regurgitation in the postoperative period, which remains the most important factor of postoperative morbidity and mortality [18]. Other risk factors for surgical repair include the patient's age at the time of surgery, the severity of preoperative common AV valve regurgitation, the presence of associated cardiac anomalies, and the degree of functional disability [19].

Since the first documented description of atrioventricular septal defect correction in 1954 [20], various techniques have been proposed for its repair. The main difference between them is in using either a single or double patch technique in closing the septal defects. Both these techniques seem to be equally efficacious [21]. As always, the surgeon's experience with a particular technique and the ability to adapt to the highly variable pathologic abnormality of the CAVSD is probably more important than just the technique itself.

It would, however, be desirable to adopt a simpler and reproducible technique that, with some customization, can be used consistently from one patient to another. By customizing the repair to the size and configuration of each patient's ventricular defect, we occasionally avoid a patch altogether with direct closure, use as small a patch as possible, or use a combination of these techniques. This greatly simplifies and expedites the operation. Moreover, improvements in anesthetic and intensive care have made it a relatively safe procedure. These patients are also being referred at a younger age than previously and, consequently, before the onset of unmanageable pulmonary hypertension.

In conclusion, we have found no difference in operative mortality and early morbidity in children undergoing surgical repair of CAVSD at 3 months of age or younger. We believe this to be the result of all of the factors we have discussed. In the current era, using the management strategies we have described, early repair of CAVSD is safe and effective.

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DISCUSSION

DR FRED A. CRAWFORD, JR (Charleston, SC): Dr Singh, I enjoyed your presentation and also appreciated the opportunity to review your manuscript. I congratulate you on your good results, but I am still not sure that I exactly understand the customized surgical approach that you have employed. I agree with earlier surgical intervention in these patients and believe that this has essentially eliminated the problem of postoperative pulmonary hypertension. This, combined with blood cardioplegia, has led to markedly decreased operative mortality in many series. For example, we have had no operative deaths in over 100 patients in the last 10 years with this particular defect. However, not all problems have been eliminated. Reoperation for mitral or left AV valve regurgitation remains a problem and occurs in 5% to 10% of every series reported in the literature regardless of the one-patch or two-patch technique, whichever one is employed.

I have several questions. In the manuscript, you report that only about 50% of your patients have absent or mild residual left AV valve regurgitation. Does this mean then that 45% of your patients have either moderate or severe MR on follow-up? Secondly, do you think that residual MR is related to age, size, or to the surgical technique? And then finally, based upon the above, what modifications, if any, have you considered to decrease the incidence of postoperative mitral regurgitation, which is the remaining problem affecting the long-term outcome in patients undergoing AV canal repair? Thank you, and again, I enjoyed your presentation.

DR SINGH: I will try and answer question two first, that 50% have absent or mild atrioventricular regurgitation and what happened to the other 45%? The answer to that question is that some of those were not reported or were not documented in follow-up examinations, but most of the patients did not have severe mitral valve regurgitation. It was documented in the later days, which was different from what was being reported earlier. So there is some data there that is not absolute, but rather than to assume that the unreported ones had absent AV valve regurgitation, we only reported those that we were sure of.

The other question was how we can improve postoperative mitral regurgitation or if there is any surgical technique that can be employed in the customized surgical approach. These answers require a highly skilled technical surgeon and I will defer that to Dr Irving Kron but give you my humble answer from the literature that I have read and discussions with Dr Kron. The customized approach really is with the ventricular septal defect. Sutures are placed if the defect is small and can be closed without a patch, and if it cannot be closed without a patch, then we employ a patch. Sometimes we use both those techniques. And then the rest of the procedure goes forward as a two-patch technique.

As for how we can reduce left atrioventricular valve regurgitation and mitral valve regurgitation, I think closing the cleft in the left atrioventricular valve would be an important step and maybe placing an additional suture there if one feels it is required intraoperatively. However, I believe that Dr Kron may have a better answer to that question.

DR KRON: Fred, just like you, I think our mortality has certainly improved over the last 10 years. We have gotten better at these

operations. Previously with mitral regurgitation, before we used routine TEE, we did the best we could before we left the operating room. We would now not hesitate if we saw more than trace mitral regurgitation to reopen, look at the valve and put a stitch or two in. One of the critical things is not to cause mitral stenosis; in fact, that is probably worse than mitral regurgitation. It is a fine line in these delicate valves. I think we have gotten better at this by using the results of perioperative TEE in determining what our mitral valve looks like. I can't recall a reoperation in the last four years for this issue.

DR ROSS M. UNGERLEIDER (Portland, OR): That was a nice study, and I think it underscores the point that was being made earlier in this afternoon's session by Karl Welke that the outcomes for congenital heart surgery, especially if we use mortality as the outcome parameter, seem to be very good and getting better. And so the question that I have for you really revolves around what should we be looking at to answer the question of safe and effective. Obviously the ultimate safety is survival, but there are other factors you didn't tell us about and maybe this is an opportunity that you could tell us a little bit about the morbidity differences between the early group and the late group. Was there a difference in ICU stay and other elements that might impact on long-term survival and of course the real concern that we have of operating on particularly young children with respect to long-term neurologic outcome?

On the other end of that spectrum you have your group B, that is the older child, going back to 1990, and I am wondering what the ages were in that group, because you mentioned that there were no pulmonary artery bands. And so did you have a group there that was not very dissimilar from your young group, that is, they were four or five months old, and I am not sure you are comparing things that are that dissimilar, or if that group B was a much older group, then I am curious how you got by without banding them, because some of them would probably have some pretty significant pulmonary resistance issues?

It finally gets us back to when should we operate on patients with AV canals, especially if we look at the factors beyond mortality that are the true outcome elements that tell us what we should be doing to get the optimal outcome. This is a very nice study and certainly makes us confident that we are doing pretty well in this field.

DR SINGH: As far as group B and the age range goes, I know I gave you a large range, but the mean was fairly close to around five to six months. So even though some of our patients looked like they were much older, and you are absolutely right, how they got away without PA banding, they had significant pulmonary resistance but did not require PA banding at the time. They had come back to clinic repeatedly with some pneumonia, some failure to thrive, but had got by. I believe 20% of people do get to live past a certain age to their first decade of life. But most of our patients actually were around the sort of late months to early year age.

As far as morbidity, no, we did not look at ICU stay. The only things I did get to look at were postoperative infections, pneumonia, or pleural effusion, and the results were fairly similar in both groups.