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Anatomic considerations in the management of complete atrioventricular canal

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Abstract

Objective: Patients with complete atrioventricular canal have a variable clinical course prior to repair. Many patients balance their circulations well prior to elective repair. Others manifest clinically significant pulmonary over circulation early in life and require either palliative pulmonary artery banding or complete repair. The objective of this study was to assess anatomic features that impact the clinical course of patients. *Methods:* In total, 222 patients underwent complete atrioventricular canal repair between 2012 and 2022 at a single institution. Twenty-seven (12%) patients underwent either pulmonary artery banding (n = 15) or complete repair (n = 12) at less than 3 months of age (Group 1). The remaining 195 (88%) underwent repair after 3 months of age (Group 2). Patient records and imaging were reviewed. *Results:* The median post-operative length of stay following complete repair was 25 [7,46] days for those patients in Group 1 and 7 [5,12] days for those in Group 2 ($p < 0.0001$). There was relative hypoplasia of left-sided structures in Group 1 versus Group 2. Mean z-score for the ascending aorta was −1.2 (±0.8) versus −0.3 (±0.9) ($p < 0.0001$), the aortic isthmus was −2.1 (±0.8) versus −1.4 (±0.8) ($p = 0.005$). The pulmonary valve to aortic valve diameter ratio was median 1.47 [1.38,1.71] versus 1.38 [1.17,1.53] (p 0.008). *Conclusions:* Echocardiographic evaluation of the systemic and pulmonary outflow of patients with complete atrioventricular canal may assist in predicting the clinical course and need for early repair vs pulmonary artery banding.

Patients with complete atrioventricular canal have been surgically managed for several decades with a high degree of success. Modern outcomes for operative management report mortality of 2–5%[1,2] and freedom from atrioventricular valve reoperation to be 80–90% at 10 years.[2–6] These excellent outcomes are in large part due to the extensive study that has been performed over the years to optimise outcomes for these patients. Investigations have included focused evaluation of the surgical techniques utilised. In particular, the management of the left atrioventricular valve cleft[7–10] and number of patches to be utilised for septation.[11,12] Additionally, there have since been a number of studies looking at the timing of repair and the application of pulmonary artery banding for palliation in infancy prior to complete repair. The results vary with regard to impact on mortality and impact on reoperation.[13–17]

Patients with complete atrioventricular canal defects have a variable clinical course prior to complete repair. Many balance their pulmonary and systemic circulations well and are discharged home with little to no medical management prior to elective, complete repair. Others manifest clinically significant heart failure early in life and require either palliative pulmonary artery banding or medical management with prolonged hospitalisations prior to early, complete repair. The anticipated clinical course of these patients in the neonatal and infantile period has been difficult to predict. This poses significant challenges both for management and for family counselling regarding surgical planning of their child's cardiac anatomy.

The objective of this study was to assess anatomic features on echocardiogram that could be used to anticipate the clinical course of patients with complete atrioventricular canal in order to assist in medical management, surgical strategy, and family counselling.

Materials and methods

We retrospectively identified and reviewed all patients with complete atrioventricular canal who underwent repair at the Children's Hospital of Los Angeles from 2012 through 2022. There were a total of 222 over this 10-year period. Patients with partial, transitional, and unbalanced atrioventricular canal were excluded. In addition, patients with a concomitant diagnosis of tetralogy of Fallot were also excluded from the study. There is no formalised institutional protocol to decipher management strategies of patients who are unable to be discharged home prior to elective complete atrioventricular canal repair. Preference is to achieve complete repair

Table 1. Demographics

	All n = 222	Group 1 n = 27	Group 2 n = 195	
	IQR/(%)	IQR/(%)	IQR/(%)	*p* value
Gestational age (wks)	37 [36,38]	38 [36,39]	37 (36-38)	0.11
Premature birth (%)	60 (27)	6 (22)	53 (27)	0.651
Birthweight (kg)	2.95 ± 0.66	3 ± 0.73	2.94 ± 0.64	0.473
T21 (%)	184 (83)	21 (78)	164 (84)	0.768
Male (%)	102 (46)	13 (48)	90 (46)	0.839
Non-cardiac abnorm (%)	40 (18)	9 (33)	31 (16)	0.035

IQR = interquartile range; T21 = trisomy 21; wks = weeks.

when possible. However, pulmonary artery banding is pursued in the setting of confounding medical issues that preclude candidacy for cardiopulmonary bypass.

In order to elucidate anatomic differences in patients that underwent early versus late repair, we separated our cohort into two groups. Group 1 had either pulmonary artery banding or complete repair at < 90 days of age. Group 2 underwent complete repair at > 90 days of age. In Group 1, there were twenty-seven (12%) patients with fifteen undergoing pulmonary artery banding and twelve with complete repair. Group 2 included the remaining 195 (88%) patients. We collected demographic, perioperative, echocardiographic data, and mortality through the time of complete repair.

The first echocardiogram of each patient was reviewed to measure the left ventricular outflow tract structures. The structures measured included the aortic valve annular area, ascending aorta diameter (at the level of the right pulmonary artery), distal transverse arch diameter (distal to left carotid and proximal to left subclavian), and aortic isthmus (distal to left subclavian). In addition, the ventricular septal defect depth was measured in four-chamber view from the crest of the septum to the plane of the overlying atrioventricular valve in end systole. Z-scores for these structures were calculated using body surface area at the time of the imaging and the Boston Z-score calculator (https://zscore.chboston.org/). This project was approved by the Children's Hospital of Los Angeles Institutional Review Board (#CHLA-22-00371; approved 4/6/23).

Statistical analysis

Data were analysed with SAS 9.4. Normally distributed continuous data is reported as mean with standard deviation while non-normally distributed data is presented as median with interquartile range. Categorical data are expressed as percentages. Comparisons of normally distributed continuous variables were done using a t-test, and comparisons of non-normally distributed variables were done using Mann Whitney U test. In each case, significance was defined as $p < 0.05$. Receiver operating characteristic (ROC) curve analysis of assignment to early versus elective complete atrioventricular canal repair was carried out to determine an inflection point of pulmonary valve to aortic valve diameter ratio. A confidence interval of 95% was utilised. This was confirmed with Youden statistic.

Results

Two hundred and twenty-two patients underwent complete atrioventricular canal repair at our institution during the study period. For this group, the median gestational age at birth was 37 (interquartile range 36–38) weeks. Twenty-seven per cent were born premature (<36 weeks). The mean weight at birth was 2.95 (SD ± 0.66) Kg. Eighty-three per cent were trisomy 21. Forty-six per cent were male and nearly a fifth had non-cardiac abnormalities. The only demographic parameter that demonstrated statistical significance between the early repair/pulmonary artery banding group (Group 1) and the late repair group (Group 2) was the number of non-cardiac abnormalities 33% versus 16% ($p = 0.035$), respectively. (Table 1)

Median age and weight at the time of complete repair for the overall cohort were 189 [153,220] days and 5.6 [4.8,6.5] Kg. Median cardiopulmonary bypass and aortic cross-clamp times were 91 [71,110] min and 71 [54,87] min, respectively. Median hospital length of stay and post-operative length of stays were 8 [5,17] and 7 [5,15] days. The overall hospital mortality for patients undergoing complete atrioventricular canal repair was 1.8%. Group 1 patients were younger 146 [75,218] versus 189 [157,220] days at the time of complete repair ($p = 0.013$). The median age at the time of complete repair for those who previously underwent pulmonary artery banding was 216 (175, 310) days. The median age at the time of complete repair for those in Group 1 who did not have a pulmonary artery banding was 70 (54,79) days. (Supplemental Table 1) The hospital length of stay for Group 1 was four times longer ($p < 0.0001$) and the post-operative length of stay was over three times longer ($p < 0.0001$) than Group 2. The median post-operative length of stay following complete repair was similar between those patients in Group 1 that had previous pulmonary artery banding performed and those without previous pulmonary artery banding; 24 (7,71) and 29 (8,35) days, respectively ($p = 0.97$). In addition, there was an increased mortality in Group 1 that reached statistical significance. (Table 2)

The median age and mean weight at the time of pulmonary artery banding were 35 [20, 55.5] days and 3.59 (±0.64) kg. Median post-operative length of stay following pulmonary artery banding was 30 [17,77.5] days and hospital length of stay was 42 [24,105] days. A total of 5/15 (33%) had pulmonary artery banding performed at the time of neonatal intervention for aortic arch augmentation. There were 11 major post-operative complications[18] in the pulmonary artery banding patients with no in-hospital mortality. One patient in this group did not survive to 30 days following return for complete repair, secondary to airway complication at home. Two banded patients required operative reintervention for adjustment prior to complete repair. A total of 3/15 (20%) of patients who underwent pulmonary artery banding remained hospitalised until complete repair was completed. Of the Group 1 patients who were discharged home prior to complete

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Table 2. Details of complete repair

	All n = 222	Group 1 n = 27	Group 2 n = 195	
	IQR/(%)	IQR/(%)	IQR/(%)	*p* value
Age (d)	189 [153,220]	146 [75,218]	189 [157,220]	0.013
Weight (kg)	5.6 [4.8,6.5]	5 [4.1,6.5]	5.6 [5,6.5]	0.074
CPB (min)	91 [71,110]	101 [78,128]	91 [56,97]	0.064
X-clamp (min)	71 [54,87]	76 [56,78]	70 [54,85]	0.164
HLOS (d)	8 [5,17]	30 [8,51]	7 [5,14]	<0.0001
Post-op LOS (d)	7 [5,15]	25 [7,46]	7 [5,12]	<0.0001
Mortality (%)	4 (1.8)	2 (7.4)	2 (1.0)	0.04

D = days; HLOS = Hospital length of stay; IQR = interquartile range; LOS = Length of stay.

repair, 33% (4/12) were able to achieve full oral feeding. The remaining 67% (8/12) were discharged home with supplemental or full enteral feeds via either home nasogastric tube or surgical gastric tube. Diuretic therapy was continued at discharge in 92% (11/12) and ace inhibitor therapy in 42% (5/12).

The median age and mean weight at the time of early canal repair was 70 [54,79] days and 3.87 (±0.64) kg. Median post-operative length of stay following early canal repair was 28.5 [7.75,35] days and hospital length of stay was 35.5 [8.8,51.3]. There were nine major complications and one mortality in this group prior to discharge. This child had a history of duodenal atresia status post repair and experienced an abdominal catastrophe after canal repair from which they ultimately were unable to recover. Of the patients that survived to discharge, 82% (9/11) were able to achieve full oral feeding. Eighteen per cent (2/11) required supplemental or full enteral feeds via either home nasogastric tube or surgical gastric tube at the time of discharge. Diuretic therapy was continued at discharge in 64% (7/11) and ace inhibitor therapy in 45% (5/11).

Overall, we found the systemic outflows in Group 1 to be smaller than those in Group 2 on the first echocardiogram obtained for each patient. The ratio of the diameter of the pulmonary valve to the diameter of the aortic valve was found to be statistically significantly larger in Group 1 as compared to Group 2, 1.47 [1.38,1.71] versus 1.38 [1.17,1.53] ($p = 0.008$). Both the mean diameter and z-score of the ascending aorta were smaller in Group 1 as compared to Group 2, 6.8 ± 1.1 mm versus 8.8 ± 1.7 ($p < 0.0001$) mm and -1.2 ± 0.8 versus -0.3 ± 0.9 ($p < 0.0001$). In addition, the mean diameter and z-score of the aortic isthmus were smaller in Group 1 as compared to Group 2, 3.4 ± 0.8 versus 4.6 ± 1.1 ($p = 0.001$) and -2.1 ± 0.8 versus -1.4 ± 0.8 ($p = 0.003$). There was no statistically significant difference in the depth of the VSD, transverse aortic arch z-score, or degree of atrioventricular valve regurgitation between the two groups (Table 3).

Receiver operating characteristic curve analysis of assignment to early versus elective complete atrioventricular canal repair (AUC 0.666, 95% CI 0.5485,0.7482) has demonstrated an inflection point of 1.37 for pulmonary valve to aortic valve diameter ratio. (Fig. 1)

Table 3. First echocardiogram anatomic measurements

	Group 1 n = 27	Group 2 n = 195	
	IQR	IQR	*p* value
Pulmonary/aortic valve ratio	1.47 [1.38,1.71]	1.38 [1.17,1.53]	0.008
	Mean Diameter (mm)	Mean Diameter (mm)	
Ascending aorta	6.8 ± 1.1	8.8 ± 1.7	<0.0001
Distal transverse arch	4.2 ± 1.3	5.4 ± 1.5	0.014
Aortic isthmus	3.4 ± 0.8	4.6 ± 1.1	0.001
VSD depth	7.5 ± 1.7	7.8 ± 2.6	0.580
	Z-Score	Z-Score	
Ascending aorta	−1.2 ± 0.8	−0.3 ± 0.9	<0.0001
Distal transverse arch	−2.1 ± 1.2	−1.5 ± 1.1	0.129
Aortic isthmus	−2.1 ± 0.8	−1.4 ± 0.8	0.003
AVVR grade	%	%	
None	33%	22%	0.223
Mild	11%	49%	
Moderate	45%	26%	
Severe	11%	3%	

AVVR = Atrioventricular valve regurgitation; IQR = interquartile range.

Discussion

Our institutional approach to patients with complete atrioventricular canal has been to perform repair at 4 months of age and 4 kilograms of weight when patients' clinical progress allows. This strategy has been supported by the 2014 Society of Thoracic Surgeons Congenital Heart Surgery Database study which reported an in-hospital mortality for children undergoing complete atrioventricular canal repair of 9.5% for children younger than 2.5 months of age and 15.2% mortality for those weighing less than 3.5 kg.[19]

The goal is that patients will be admitted on an elective basis from home for repair. As a general rule, our patients then typically undergo repair with two patches and primary closure of the left atrioventricular valve cleft. If a child is unable to wean from respiratory support or demonstrates poor weight gain on enteral feeds, while receiving maximum oral medical therapy, we consider performing either pulmonary artery banding or early (<3 months) complete repair.

As our data demonstrates, among all neonates presenting with complete atrioventricular canal, we perform early repair (<90 days) or pulmonary artery banding 12% of the time. Though

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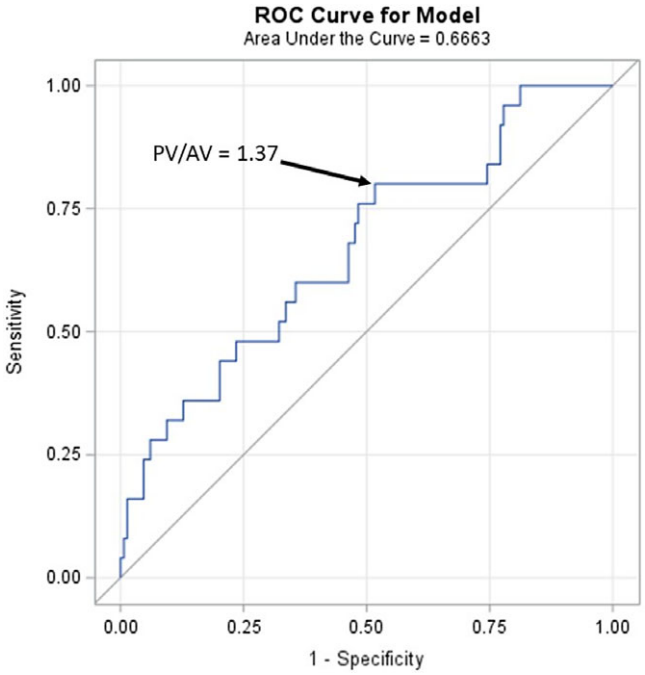

Figure 1. Receiver operating characteristic curve.

this population includes a small percentage of our overall experience with complete atrioventricular canal repair, the hospital length of stay, post-operative length of stay, and mortality of these patients are significantly different from those that undergo elective repair. Our median post-operative length of stay for the overall cohort was 7 [5,15] days compared with the Society of Thoracic Surgeons Congenital Heart Surgery Database median post-operative length of stay most recently reported to be 17.9 [13.3,23.6] days.[20] However, the median post-operative LOS for the elective repair group was 7 [5,12] versus 25 [7,46] days for the early/pulmonary artery banding group. These findings mark a contrast in the anticipated post-operative course that could help with counselling with family and the care team about appropriate expectations.

Our data has shown a statistically significant difference in the size of the systemic outflow on initial echocardiogram in Group 1 as compared to Group 2. The ascending aorta and aortic isthmus were smaller in Group 1 in both indexed and non-indexed measurements. In addition, the pulmonary valve to aortic valve diameter ratio was found to be larger in Group 1. We hypothesise that the smaller left-sided structures cause an increase in left to right shunting with over circulation of the pulmonary bed secondary to the cumulative resistance caused by the relative hypoplasia of the aortic valve and aorta as compared to the pulmonary outflow. We believe that this induced pulmonary over circulation causes the clinical manifestations of heart failure that prompt consideration of either pulmonary artery banding or early complete canal repair.

Hypoplasia of the systemic outflow in complete atrioventricular canal poses a potentially difficult anatomy to balance systemic and pulmonary circulations with pulmonary artery banding. This difficulty in balancing circulations secondary to hypoplastic systemic outflow may contribute to an unstable circulation in the interstage period prior to complete repair. An adequate balance may induce physiology favouring either pulmonary over circulation or bilateral outflow tract obstruction. A recently published extensive retrospective review of complete atrioventricular canal patients undergoing either pulmonary artery banding or complete repair at less than 3 months of age reported an 18.6% mortality during the interstage period prior to complete repair.[21] Though we did not experience mortality in the interstage period, review of details of Group 1 patients at hospital discharge shows that only 33% of those who underwent banding were able to achieve full oral feeds as compared to 82% of those who underwent complete repair. Also, 92% of pulmonary artery banding patients were discharged on diuretic therapy as compared to only 64% of those in the complete repair group. These findings support evidence of continued pulmonary over circulation in our pulmonary artery banding patients at discharge.

Our data has demonstrated the utility of echocardiographic assessment of the systemic and pulmonary outflow anatomy in patients with complete atrioventricular canal. Patients diagnosed with complete atrioventricular canal should undergo full transthoracic echocardiographic assessment of the systemic outflow to include measurements of the aortic valve annulus, ascending aorta, transverse arch, and aortic isthmus, as well as imaging of the right ventricular outflow tract to assess for evidence of sub- and supravalvar right ventricular outflow obstruction and specific measurement of the pulmonary valve annulus. Receiver operating characteristic curve analysis has indicated that patients with pulmonary valve to aortic valve diameter ratio > 1.37 are likely to require early intervention secondary to clinical heart failure with 67% accuracy. This information can then be utilised to more accurately counsel family and care teams regarding the anticipated clinical course of these patients.

Limitations

This is a single institution retrospective non-randomized experience regarding management of complete atrioventricular canal. Therefore, the decision to perform a pulmonary artery band versus complete repair and the resultant timing is subject to institutional and individual surgeon bias. The retrospective nature of this study also resulted in incomplete data.

Conclusions

Echocardiographic evaluation of the systemic and pulmonary outflow of patients with complete atrioventricular canal may assist in predicting the clinical course and need for early repair versus pulmonary artery banding. Echocardiographic findings of hypoplastic systemic outflow portend a need for early intervention to address pulmonary overcirculation. These anatomic findings on imaging should be used to counsel caregivers and families as to the likely clinical course and need for early intervention.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951123003323.

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Competing interests. None.

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