

Risk factors for adverse outcomes after surgery on the systemic atrioventricular valve in 109 children

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IN CONTRAST TO OLDER PATIENTS, CHILDREN AND young adults rarely have isolated disease of the systemic atrioventricular valve. Stenosis and/or regurgitation of the systemic atrioventricular valve, however, frequently coexist with complex congenital cardiac disease. In addition, most patients undergoing surgery on the systemic atrioventricular valve have had previous intracardiac repairs.

Despite the heterogeneity of the underlying anatomy, physiology, and co-morbidities related to the patient, repair or replacement of the systemic atrioventricular valve in all cases acutely changes the loading conditions of the myocardium, producing decreased preload and increased afterload. In cases of valvar regurgitation, the effect of the acute change in load on postoperative ventricular mechanics depends in large part on the preoperative functional state of the systemic ventricle. Preoperative ventricular decompensation, as manifest by progressive dilation, decreased ejection fraction, and low cardiac output, may exaggerate the expected postoperative and post-ischaemic decline in ventricular function following repair or replacement of the valve, resulting in low cardiac output. In cases of valvar stenosis, elevated preoperative pulmonary arterial pressure and vascular resistance may make postoperative pulmonary hypertensive crises more likely to occur. Multiple studies have described the postoperative challenges associated with surgery to the systemic valve in subgroups of patients with congenital cardiac disease, such as ventricular dysfunction, low cardiac

output, arrhythmias, and secondary pulmonary hypertension.^{1–6} Risk factors that predict the adverse postoperative course, however, have not yet been identified.

The goal of our study, therefore, was to identify the frequency of, and risk factors for, adverse outcomes in the early postoperative period following surgery to the systemic atrioventricular valve in children.

Materials and methods

Design of the study

We conducted a retrospective analysis, focusing on early surgical results, which was performed following approval from our Institutional Review Board. We included all patients at the Children's Hospital of Philadelphia who underwent surgery primarily for repair or replacement of the systemic atrioventricular valve between January of 1995 and April of 2003, including those with functionally single ventricles. We chose to use the term systemic atrioventricular valve so as to include the morphologically tricuspid valve in complex congenital cardiac malformations in which the morphologically right ventricle was the systemic ventricle, and also the common atrioventricular valves in patients with functionally univentricular hearts. Pre-operatively, all patients were considered to have haemodynamically significant valvar regurgitation, stenosis, or both. Repair or replacement of the valve was performed at the discretion of the surgeon. We excluded patients undergoing primary repair of atrioventricular septal defect with common atrioventricular junction, irrespective of whether or not surgery was performed on the left atrioventricular valve. We did include patients if, following repair of atrioventricular septal defect, they returned for surgery on the left-sided

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atrioventricular valve. For the purposes of this review, the term "mitral" annulus will be used, to combine measurements made on a native mitral valve, as well as those on the left component of a repaired common atrioventricular valve into one category, as they are both associated with a morphologically left ventricle, and both function as the systemic atrioventricular valve.

We reviewed the inpatient charts for age at surgery, weight, gender, anatomic diagnosis, indication for surgery, and prior interventions. The preoperative transthoracic and intraoperative transesophageal echocardiograms were reviewed for qualitative assessment of ventricular function, systemic atrioventricular valvar regurgitation, and stenosis. In patients with concordant atrioventricular connections, and two functional ventricles, measurements were made off-line to determine the preoperative area of the mitral valve, and the volume and mass of the left ventricle. The annulus of the mitral valve was measured in 2 orthogonal planes so as to calculate the valvar area. The thickness of the left ventricular free wall and septum were measured in diastole from a parasternal long-axis view, and combined with measurements of ventricular volume to determine the ventricular mass. All measurements were performed by a single observer in a blinded fashion, and converted to z scores for comparison. Where applicable, the records of preoperative cardiac catheterization were reviewed to establish the cardiac index, pulmonary arterial pressures, pulmonary vascular resistances, and end-diastolic ventricular pressures. We reviewed the operative notes and records of anaesthesia to establish the surgical procedure carried out, that is repair or replacement, any concomitant surgical procedure(s), the duration of cardiopulmonary bypass and circulatory arrest, if the latter was used, the total support time, and the number of intraoperative revisions. Intraoperative transesophageal echocardiography was performed in all patients in the operating room upon discontinuation of cardiopulmonary bypass.

Definitions

An adverse postoperative outcome was defined as death, and/or the need for mechanical circulatory support such as extracorporeal membrane oxygenation or a left ventricular assist device.

The systemic atrioventricular valve was defined as the valve contained within the systemic ventricle, and subjected to the systemic afterload.

Systemic atrioventricular valvar regurgitation was quantified by colour Doppler interrogation and graded on a scale of 0 to 4, with zero representing no regurgitation, and 1 through 4 representing a trace of, mild, moderate, or severe regurgitation.

Systemic atrioventricular valvar stenosis was determined by continuous wave Doppler interrogation of the valvar inflow. Significant stenosis was defined as a mean gradient greater than or equal to 8 millimetres of mercury.

A significant concomitant surgical procedure was defined as an additional procedure performed at the time of atrioventricular valvar surgery that could have directly altered the ventricular loading conditions, such as closure of a residual atrial or ventricular septal defect, resection of sub-aortic stenosis, and so on.

Multiple intraoperative revisions were defined as greater than 1 intraoperative revision that required re-initiation of cardiopulmonary bypass following postoperative transoesophageal echocardiography.

The primary adverse outcome measured for the study was death, and/or the need for postoperative mechanical circulatory support. Secondary outcomes measured included: re-operation, seizure, renal failure with a level of creatinine greater than 1.5, re-intubation, length of stay, and re-admission to the hospital within 30 days of discharge.

Statistical methods

Statistical analysis was performed with Stata 8.0 analysis software (Stata Corporation, College Station, Texas). Normally distributed continuous variables were described as means plus or minus standard deviations. Non-parametric continuous variables were described as the median and range. Univariate analysis of potential risk factors for adverse outcome was performed using the Wilcoxon Rank Sum test for continuous variables, and the Fisher's Exact test for dichotomous variables. Multivariate analysis was performed using Logistic regression. A p value of greater than 0.05 was considered significant.

Results

Characteristics of the patients

We found 109 patients meeting our criteria for inclusion. They had undergone 117 procedures during the period of study. There were 54 males (49.5%), and 55 females (50.5%). The median age of the group was 5.6 years with a range of 2 months to 50 years. The frequency distribution of age at time of surgery is shown in Figure 1. The majority of the cohort, 86 or 73.5%, were evenly distributed between the ages of 1 and 18 years. At the time of surgery, there were 24 (20.5%) under the age of 1 year, and 7 (6%) over the age of 18 years. The original anatomic diagnoses are shown in Table 1. In all, 114 surgical procedures had been performed previously on 75 patients in the cohort.

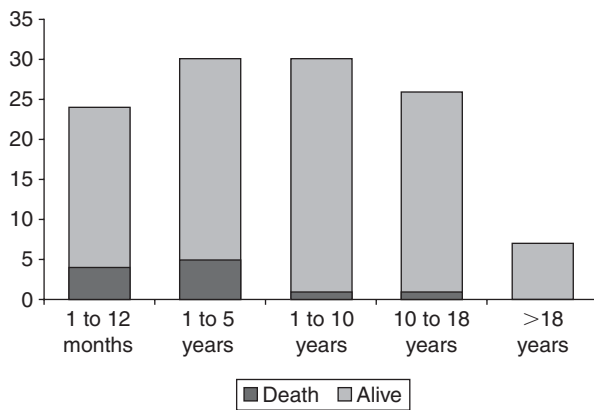


Figure 1.

Age distribution of patients undergoing surgery on the systemic atrioventricular valve, and frequency distribution of death relative to the age of the patients.

Table 1. Original anatomic diagnoses.

Preoperative anatomic diagnosis	# of patients
Secondary repair of LAVV in AVSD	43
Non-specific congenital MR	16
Arcade MV	9
Functionally single ventricle	8
Rheumatic valvar disease	7
Parachute MV	6
Isolated cleft MV	5
MV endocarditis	4
HOCM	4
Primum AVSD/"Cleft" LAVV	3
Corrected transposition	3
ALCAPA	3
Marfan syndrome	2
Transposition	1
DORV	1
Double-orifice MV	1
Intracardiac tumour	1

Abbreviation: ALCAPA: anomalous origin of the left coronary artery

from the pulmonary artery; ASD: atrial septal defect;

AVSD: atrioventricular septal defect with common atrioventricular canal;

DORV: double outlet right ventricle; HOCM: hypertrophic obstructive

cardiomyopathy; LAVV: left atrioventricular valve in atrioventricular

septal defect; Marfan: Marfan syndrome; MR: mitral regurgitation;

MV: mitral valve

Preoperative echocardiography

Preoperative transthoracic echocardiography was performed in all patients. Moderate or severe valvar insufficiency was the predominant lesion in 84 (72%). Isolated stenosis was found in 15 (13%), and 18 (15%) had a combination of significant regurgitation and stenosis. In 101 patients with functionally biventricular physiology and systemic left ventricles (93%), the mean left ventricular end diastolic diameter Z score was 1.9 plus or minus 2.6. The mean left ventricular mass Z score was 2.2 plus or minus 3.1.

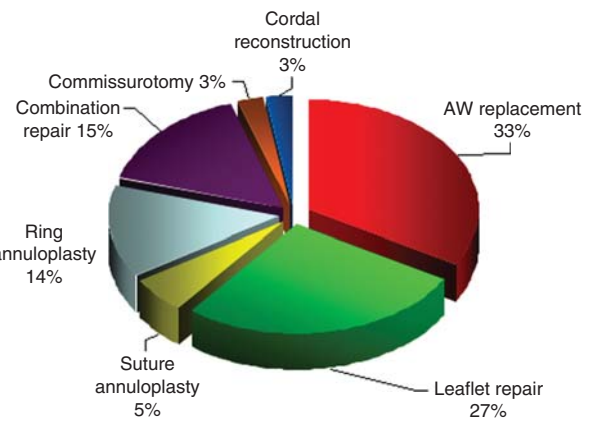


Figure 2.

Cartoon showing the details of 117 procedures, of which 78 were repairs, and 39 involved replacement of the valve.

Preoperative cardiac catheterization

There were 79 preoperative cardiac catheterizations (67.5%) obtained at a median duration prior to surgery of 18 days, with a range from 1 to 234 days. The median cardiac index was 3.2, with a range of 1.5 to 5.8 litres per minute per metre squared. The median mean pulmonary arterial pressure was 26.5 millimetres of mercury, with a range from 11 to 97. The median end diastolic pressure was 11 millimetres of mercury, and a range from 3 to 28. The median pulmonary capillary wedge pressure was 15 millimetres of mercury, the range being 4 to 28. The median pulmonary vascular resistance was 3.6 Woods units, with a range from 0.7 to 36. The pulmonary vascular resistance was greater than 4 Woods units in 36 patients (30.7%).

Operative data

During the period of study, 109 patients underwent 117 admissions for surgery on the systemic atrioventricular valve, with 78 repairs (67%), and 39 replacements (33%) (Fig. 2). Of the patients, 32 (27%) required greater than 1 intraoperative revision based on the findings from intraoperative transoesophageal echocardiography, and/or adverse haemodynamics. Additional operative procedures were performed in 51 patients (44%). In 36 patients, the concomitant procedures had potential haemodynamic significance, including closure of residual atrial or ventricular septal defects in 12, surgery to the aortic valve in 9, with the valve being replaced by a mechanical prosthesis in 5, an autograft in 2, and repaired in 2, and resection of subaortic stenosis in 7. Revision of the Fontan circulation had been undertaken in 3, surgery performed on the right ventricular outflow in 2, with the pulmonary valve being replaced in 1 and a conduit revised in the

other, and completion of the Fontan circulation, repair of the tricuspid valve, and repair of coarctation in 1 patient each. Revision or replacement of a pacemaker was performed in 14 patients, and one patient had a concomitant lung biopsy.

Perfusion data

The perfusion data is shown in Table 2. The median time of cardiopulmonary bypass was 77 minutes, with a range from 24 to 360 minutes. Circulatory arrest was used in 26 patients (22%, the median period being 38 minutes, with a range from 7 to 110 minutes. The median period of aortic cross-clamping was 46 minutes, with a range from 12 to 209 minutes. The median total support period, including cardiopulmonary bypass and circulatory arrest, was 81 minutes, with a range from 24 to 394 minutes.

Intraoperative transoesophageal echocardiography

Prior to leaving the operating room, 36 patients (30.7%) had depressed ventricular function.

Table 2. Perfusion data.

Times	Median (minutes)	Range (minutes)
Cardiopulmonary bypass	77	24–360
Circulatory arrest (n = 26)	38	7–110
Aortic cross-clamp	46	12–209
Total support	81	24–394

Significant residual systemic atrioventricular regurgitation was noted in 17 of them (14.5%), and significant residual stenosis, with a mean gradient greater than or equal to 8 millimetres of mercury, was found in 8 (6.8%).

Mortality and postoperative mechanical circulatory support

A total of 16 patients (13.7%) suffered an adverse outcome, with 11 postoperative deaths (9.4%), 7 of whom also had postoperative circulatory support. An additional 5 patients underwent postoperative circulatory support and survived (Table 3). The median age of those who died was 2.3 years, with a range from 2 months to 15 years. The frequency of distribution of death relative to the day of admission to the Intensive Care Unit is shown in Figure 3. The median duration of extracorporeal membrane oxygenation was 144 hours, with a range from 2 to 744 hours. The patient treated with a left ventricular assist device died on the sixth postoperative day. Of the 8 patients who had functionally univentricular physiology, 4 underwent repair of the systemic atrioventricular valve, with the valve being replaced in 4. None of the patients with a functionally single ventricle required mechanical circulatory support, and none died.

Significant risk factors for death and/or mechanical circulatory support

In a univariate analysis, multiple risk factors were identified for adverse outcomes, many of which were

Table 3. Mortality and postoperative mechanical circulatory support.

Patient	Death	ECMO/ LVAD	Age	Diagnosis	Valve surgery	PVR*	>1 inraop revision	Depressed function on TEE
1	+	–	4m	AVSD/DORV/PS s/p MBTS	Repair	4.6	+	+
2	+	–	1.5y	Arcade MV	Replace	5.2	+	+
3	+	–	2m	HOCM	Replace	6	–	+
4	+	–	3y	TGA s/p Rastelli	Replace	n/a	+	+
5	+	+	6y	AVSD	Repair	10	+	+
6	+	+	2m	Parachute MV	Replace	5.7	+	+
7	+	+	2y	Arcade MV	Replace	5	+	+
8	+	+	3m	Arcade MV	Replace	11	+	+
9	+	+	4.5y	Isolated cleft MV	Repair	n/a	–	+
10	+	+	4y	Marfan	Replace	n/a	+	+
11	+	+	15.3y	Arcade MV	Replace	36	–	–
12	–	+	2m	Parachute MV	Repair	14	–	–
13	–	+	5y	Unbalanced AVSD s/p stg 1	Repair	1	–	+
14	–	+	1.5y	Primum AVSD	Replace	0.9	+	–
15	–	+	13y	Rheumatic disease	Repair	20	+	+
16	–	+	5m	Shone's syndrome	Replace	9.6	–	+

* Preoperative PVR reported in Woods units; n/a = not available

Abbreviations: MV: mitral valve; AVSD: atrioventricular septal defect with common atrioventricular canal; DORV: double-outlet right ventricle; PS: pulmonary stenosis; MBTS: modified Blalock-Taussig shunt; HOCM: hypertrophic obstructive cardiomyopathy; AS: aortic stenosis; TGA: transposition of the great arteries; VSD: ventricular septal defect; replace: SAVV replacement; repair: SAVV repair

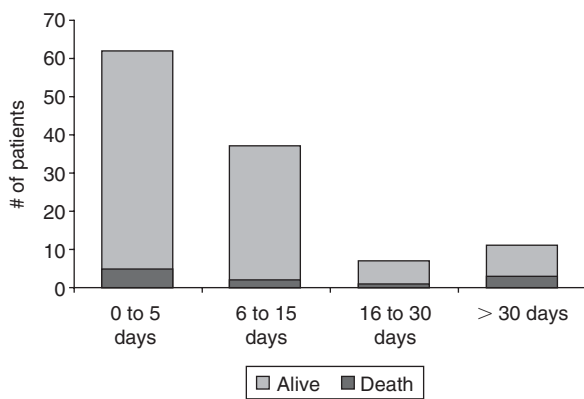


Figure 3.

Frequency distribution of death relative to postoperative day. The X-axis represents hospital length of stay.

co-linear. On multivariate analysis, significant risk factors for death and/or mechanical support included: younger age at surgery (p is equal to 0.037), higher preoperative pulmonary vascular resistance (p is equal to 0.012), more than 1 intraoperative revision (p is equal to 0.045), and depressed ventricular function as identified at intraoperative transoesophageal echocardiography (p is equal to 0.009) (Table 4). In Figure 4, we show the frequency distribution of death relative to preoperative pulmonary vascular resistance.

Postoperative morbidity

During initial hospitalization, 21 patients (17%) underwent reoperation after initial return to the Cardiac Intensive Care Unit. Of these, 8 required

Table 4. Risk factors for postoperative death and /or ECMO/LVAD.

Variable		P value	
		Univariate	Multivariate
Demographic data	Younger age	0.01	0.037
	Male sex	NS	–
	Lower weight	0.009	–
	Lower BSA	0.01	–
	Presence of preoperative symptoms	0.04	–
Anatomic diagnosis	Prior surgery	NS	–
	AVSD	NS	–
	MV arcade	0.03	–
	Parachute MV	0.04	–
	Prosthetic valve	NS	–
	Isolated MV cleft	NS	–
	Functionally single ventricle	NS	–
	Rheumatic disease	NS	–
Preoperative ECHO	Congenital MR	NS	–
	Other	NS	–
	Larger LVEDd Z score	NS	–
	Larger LV mass	0.01	–
	Larger LV mass Z score	NS	–
	Lower shortening fraction	NS	–
	SAVV regurgitation	NS	–
Preoperative catheterization	SAVV stenosis	NS	–
	Combined regurgitation/stenosis	NS	–
	Lower cardiac index	NS	–
Operative data	Higher PVR	0.02	0.012
	Longer CPB	0.003	–
	Longer aortic cross-clamp	0.004	–
	Longer DHCA	0.005	–
	Longer TST	<0.001	–
	Valve replacement	0.005	–
	Multiple revisions	0.004	0.045
	Significant concomitant procedure	NS	–
TEE data	Residual SAVV regurgitation	NS	–
	Residual SAVV stenosis	NS	–
	Depressed function	0.001	0.009

*Statistical significance defined as P value ≤ 0.05 .

Abbreviations: BSA: body surface area; AVSD: atrioventricular septal defect with common atrioventricular canal; MR: mitral regurgitation; LVEDd: left ventricular end-diastolic dimension; LV: left ventricle; SAVV: systemic atrioventricular valve; PVR: pulmonary vascular resistance; CPB: cardiopulmonary bypass time; DHCA: deep hypothermic cardiac arrest time; TST: total support time

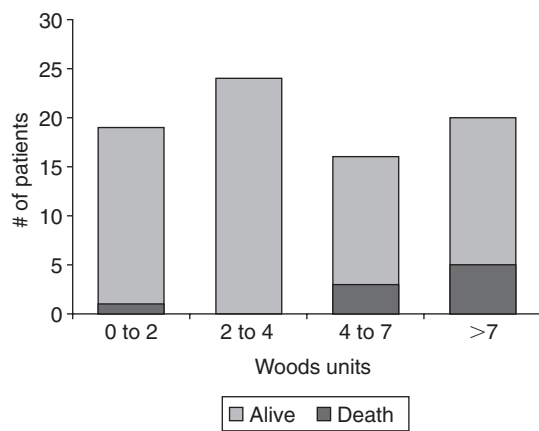


Figure 4.

Frequency distribution of postoperative death relative to preoperative pulmonary vascular resistance (PVR). Preoperative catheterization data available for 79 of 117 admissions.

surgical revision of the valve, 6 were re-explored for persistent bleeding, 2 underwent placement of a pacemaker for complete heart block, 2 underwent cardiac transplantation following extracorporeal membrane oxygenation, 1 required plication of the left hemi-diaphragm, and 1 required coronary arterial bypass surgery. Re-intubation was required in 11 patients (9.4%), 8 (6.8%) had new onset of seizure activity, 4 (3.4%) had biochemical evidence of renal failure, and 9 (7.7%) were re-admitted to the hospital within 30 days of discharge. No risk factors related to the patient or the procedure were significantly associated with these postoperative events.

Discussion

In this study, we reviewed our experience with surgery on the systemic atrioventricular valve, seeking to identify risk factors for postoperative death and mechanical circulatory support. The group of patients investigated is at high risk for such events, with just over one-eighth dying, and/or requiring mechanical support, in the early postoperative period. Mortality was high at 9.4%, approaching that for the first stage of Norwood reconstruction for hypoplastic left heart syndrome,⁷⁻⁹ and there was considerable morbidity, including two transplantations, resulting in increased consumption of resources in the intensive care unit.

The surgical literature yields variable results in this population. Most studies focus on surgery to the mitral valve, with a reported mortality from 2.2 to 30%.^{1,3,5,10,11} When it is necessary to replace the mitral

valve, rates of death may be even higher, ranging from 18 to 50%, with younger patients being particularly at risk.¹²⁻¹³ Early postoperative death following such surgery is most often attributed to progressive ventricular dysfunction, pulmonary hypertension, and complete heart block.³⁻⁴ Significant risk factors, including Shone's syndrome, the arcade lesion, age less than 1 year, increased periods of cardiopulmonary bypass, the need for a large prosthesis relative to the size of the patient, and an increased cardiothoracic ratio^{2,5,14} have all previously been identified, but supporting data is lacking, and the data which exists is conflicting.^{1,4} No studies have identified risk factors for postoperative mechanical circulatory support.

In our series, a total of 16 patients (13.7%) died and/or required postoperative mechanical circulatory support. Extracorporeal membrane oxygenation, or a ventricular assist device, was used in 12 (9.8%), 7 of whom died. The total number of deaths was 11, which is consistent with recently published reports.^{1,3-5,10-12,13-17}

The diagnoses and surgical procedures for those patients who had adverse outcome are shown in Tables 2 and 3. Of the 11 (45%) deaths, and 7 of the 12 (58%) who had mechanical support, 6 required replacement of the valve because of presence of either an arcade or a parachute lesion. Although this anatomic subgroup did not reach statistical significance in a multivariate analysis, it does lend support to a prior report² that this morphology puts patients at higher risk postoperatively, most likely from pulmonary hypertension and/or acute changes in ventricular loading. Most of the deaths occurred in the early postoperative period, suggesting that preoperative and intraoperative variables have a major impact on survival. In our series, younger age at surgery (p is equal to 0.037), and higher pulmonary vascular resistance (p is equal to 0.012), were both identified as significant preoperative risk factors. Other demographic, anatomic, clinical, and echocardiographic factors were not identified as risk factors, including: left ventricular mass; left ventricular end diastolic volume; ventricular shortening fraction; and severity of stenosis/regurgitation. Only two intraoperative factors, namely multiple intraoperative revisions (p is equal to 0.045); and, depressed function as identified by transoesophageal echocardiography (p is equal to 0.009), proved to be significant. Contrary to other recent reports,^{2,14} the periods required for intraoperative perfusion were not associated with death. In addition, significant concomitant surgical procedures were not associated with death or the need for mechanical circulatory support (p is equal to 0.37). None of the patients with functionally univentricular physiology died or required mechanical circulatory support. This is consistent with several studies that report

successful surgery on the atrioventricular valves, including valvar replacement, in patients with functionally univentricular hearts.^{18–20}

The results of our study show that a significant subgroup of children undergoing surgery on the systemic atrioventricular valve is at high-risk for mortality and morbidity. The acute changes in myocardial mechanics that occur postoperatively must be anticipated if these patients are to be managed optimally. Preoperative systemic atrioventricular valvar regurgitation results in increased preload and decreased afterload on the ventricle, and in some cases leads to ventricular decompensation. Surgery to restore valvar competence reduces both the regurgitant volume, and the ability of the ventricle to decompress into a low-pressure atrium. The result is a precipitous fall in preload, and an abrupt increase in afterload. In a deconditioned myocardium, ventricular failure, low cardiac output, and death can occur. Conversely, preoperative stenosis results in reduced ventricular preload, as well as poor ventricular compliance, and left atrial and pulmonary hypertension. In the absence of obstruction of the outflow tract, ventricular afterload is largely unaffected. Repair of valvar stenosis may improve valvar function, but it does not result in an immediate improvement in ventricular compliance or pulmonary vascular resistance. In addition, cardiopulmonary bypass may exacerbate pre-existing atrial and pulmonary hypertension.¹⁴ These physiologic changes are compounded in the presence of procedural complications, such as obstruction of the ventricular outflow tract, obstruction of a coronary artery, failure of the prosthesis, and heart block.

Recommendations for postoperative care

Our current approach to postoperative management of patients undergoing surgery on the systemic atrioventricular valve includes invasive monitoring of pressures using arterial and transthoracic atrial lines, serial assessment of cardiac output using arterial blood gases and measurements of lactate in the serum, along with mixed-venous saturations of oxygen if available, afterload reduction with inhibitors of phosphodiesterase such as milrinone, mechanical ventilation until stable, serial transthoracic echocardiography, nitric oxide to minimize pulmonary vascular lability in selected patients, and intravenous inotropic support in most patients. Providers of care should have a low threshold for an increased level of support in younger patients, those with elevated pulmonary vascular resistance, those that require intraoperative revision and, those that have depressed ventricular function by intraoperative echocardiography.

Conclusion

Surgery on the systemic atrioventricular valve is performed for a variety of indications in a heterogeneous population of children. Mortality, and the need for intense and/or prolonged support in the intensive care unit are considerable. Younger age at surgery, higher preoperative pulmonary vascular resistance, multiple intraoperative revisions, and depressed ventricular function on intraoperative transoesophageal echocardiogram, were all identified as predictors of both mortality and mechanical support. The intensivist, cardiologist, surgeon, and bedside staff need to know the preoperative state of the ventricle, the pulmonary vascular resistance, and the intraoperative procedures and echocardiographic findings, as these may determine the likelihood of postoperative acute deterioration/cardiac arrest, duration of afterload reduction, and the strategy for sedation. Patients identified as being at high risk may benefit from increased monitoring and prophylactic escalation of support during the early postoperative period. The postoperative clinical, laboratory and echocardiographic findings will dictate the strategy of medical management, which must be individualized.

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