

Prevention of Early Sudden Circulatory Collapse After the Norwood Operation

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Background—After modifications in our perioperative management protocol, we have observed a decrease in sudden circulatory collapse after the Norwood operation. The current study examines early outcomes after the Norwood operation in our unit in an attempt to identify variables that may have altered the risk of unexpected circulatory collapse.

Methods and Results—We studied 105 consecutive neonates who underwent a Norwood operation in our institution. Our treatment protocol has changed in the past 3 years to include the use of alpha-blockade with phenoxybenzamine (POB) for systemic afterload reduction and selective cerebral perfusion. Forty-eight infants had selective cerebral perfusion. Forty-two infants received POB. Sixty patients had hypoplastic left heart syndrome. There was no difference in age, diagnosis, number of neonates with weight <2.5 kg, aortic size diameter <2 mm, highest preoperative lactate level, and shunt size indexed to body weight among patients with or without use of POB. Twenty-five infants had circulatory collapse during the first 72 hours. Twelve of them could be explained by technical issues. Thirteen others who appeared clinically stable had early sudden circulatory collapse without an apparent cause. Sixteen out of 25 neonates died. Of those with technical problems, 8 out of 12 died. Based on the hazard function, 3 incremental risk factors for early circulatory collapse were technical issue at operation ($P<0.001$), longer cross-clamp time ($P<0.007$), and no use of POB ($P<0.002$). For a technically successful operation, freedom from circulatory collapse at 72 hours is 95% with the use of POB versus 69% without ($P<0.002$). Diagnosis, aortic size, atrioventricular valve function, birth weight, age at operation, and total circulatory arrest time and were not predictive of early sudden circulatory collapse.

Conclusion—Recent changes in our treatment protocol have resulted in a decrease incidence of sudden circulatory collapse after the Norwood operation. Optimal surgical technique is the most important predictor of early survival. The use of aggressive afterload reduction with POB reduced the risk of early sudden arrest. (*Circulation*. 2004; 110[suppl II]:II-133–II-138.)

Key Words: sudden death ■ congenital heart defects ■ risk factors

The outcomes for neonates with hypoplastic left heart syndrome and its variants have improved dramatically since the introduction of staged reconstructive surgery in the early 1980s.¹ Recent modifications in surgical technique and perioperative management have contributed to the observed improvement in survival.^{2–5} Traditional postoperative management after the Norwood operation has primarily consisted of balancing the pulmonary and systemic circulations by manipulation of pulmonary vascular resistance through judicious control of arterial oxygenation and pH.^{2–5} However, unexpected cardiovascular collapse and sudden death in a neonate who appeared clinically stable has not been an infrequent event. Large series have documented the prevalence of early postoperative mortality with up to 50% of deaths occurring in the first 48 hours.^{2,5,6}

Alpha blockade with phenoxybenzamine (POB) has been used in the postoperative management after the Norwood

operation. The principle of this approach is the increase in systemic cardiac output by maximal dilatation of the systemic circulation.^{7–9} This effect results in a more stable parallel circulation through prevention of fluctuations in systemic vascular resistance in the early postoperative period.⁹ We have incorporated this approach in our postoperative management protocol¹⁰ and have observed a decrease in sudden cardiac collapse. The current study examines early outcomes after the Norwood operation in our unit in an attempt to identify variables that may have altered the risk of unexpected circulatory collapse.

Methods

Patient Population

Between January 1996 and June 2002, 105 consecutive children underwent a Norwood operation at our institution. There were 65

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males (62%). Prenatal diagnosis was known in 37 infants (35%). Median age at operation was 7.5 days (range, 1 day to 5.8 months). Mean weight at operation was 3.3 ± 0.6 kg. Preoperative inotropic support was used in 29 (28%), and mechanical ventilation was required in 51 infants (49%). Median highest preoperative lactate was 4.7 (range, 1.2 to 29).

Morphological Characteristics

Diagnosis and morphology were established by echocardiography. Classic hypoplastic left heart syndrome was present in 60 infants: aortic stenosis/mitral stenosis, $n=22$ (37%); aortic stenosis/mitral atresia, $n=20$ (33%); aortic atresia/mitral stenosis, $n=10$ (17%); and aortic atresia/mitral atresia, $n=8$ (13%). Non hypoplastic left heart syndrome diagnoses included unbalanced atrioventricular septal defect ($n=11$), double outlet right ventricle ($n=8$), double inlet left ventricle ($n=8$), tricuspid atresia with transposed great vessels ($n=5$), and other ($n=13$).

Mean ascending aortic diameter was 4.5 ± 2.0 mm (range, 1.2 to 9.6 mm). Atrioventricular (AV) valve function was normal in 69 infants. AV valve regurgitation was present in 36 infants (mild, $n=33$; moderate, $n=3$). An aberrant right subclavian artery was present in 5 infants.

Clinical Management

From January 1996 to July 1999, we used conventional postoperative management. After July 1999, we used alpha blockade and more intensive perioperative monitoring. Further description is provided.

Operative Technique

Our surgical technique has been described elsewhere.³ Since 1999, we have used low-flow cardiopulmonary bypass (CPB) and selective cerebral perfusion to limit hypothermic circulatory arrest time. CPB is established by cannulation of the proximal end of a Gore-Tex conduit used for the right modified Blalock-Taussig shunt as described by Pigula et al.¹¹ The aortic arch is reconstructed with a homograft patch. The right modified Blalock-Taussig shunt is completed with anastomosis of the proximal conduit to the central pulmonary artery.

For the overall cohort, mean circulatory arrest time and mean aortic cross clamp time were 31 ± 22 and 55 ± 19 minutes, respectively. The mean selective cerebral perfusion time was 50 ± 17 minutes ($n=48$). Circulatory arrest time for patients undergoing selective cerebral perfusion was 9 ± 13 versus 50 ± 18 for patients who had circulatory arrest used as a primary strategy ($P < 0.001$). For patients undergoing selective cerebral perfusion the aortic cross-clamp time was longer, 66 ± 34 versus 50 ± 18 ($P=0.004$). Shunt diameter was 3.5 mm ($n=88$), 3.0 mm ($n=14$), or 4.0 mm ($n=3$). There was no difference in age, diagnosis, number of neonates with weight < 2.5 kg, aortic size diameter < 2 mm, highest preoperative lactate level, and shunt size indexed to body weight among patients with or without use of POB.

Traditional Postoperative Management (January 1996 to July 1999)

Inotropic support was initiated at the time of weaning from CPB. Usual agents were milrinone (0.66 to 0.99 $\mu\text{g}/\text{kg}$ per minute), dopamine (5 $\mu\text{g}/\text{kg}$ per minute), and epinephrine (0.01 $\mu\text{g}/\text{kg}$ per minute). Management of Qp:Qs was based primarily on adjustments of mechanical ventilation to maintain arterial oxygen saturations of 70% to 80%, arterial pCO_2 of 40 to 50 mmHg, and arterial pH of 7.35 to 7.4. When necessary, hypercarbia or nitrogen was used in combination with traditional ventilatory measures to control oxygen saturations. If needed, sodium nitroprusside was used for afterload reduction.

Current Postoperative Management (July 1999 to June 2002)

Our current strategy is to generate a balanced circulation by reduction of systemic vascular resistance and optimization of cardiac

output with the use of alpha blockade.¹⁰ POB (0.25 mg/kg) is given at initiation of cardiopulmonary bypass. Before termination of bypass, milrinone (100 $\mu\text{g}/\text{kg}$) and dopamine (5 $\mu\text{g}/\text{kg}$ per minute) are administered. Dopamine is generally stopped in the intensive care unit to decrease the heart rate. POB (0.5 to 1 mg/kg per 24 hours) and milrinone (0.66 to 0.99 $\mu\text{g}/\text{kg}$ per minute) are maintained. Optimally, a mean arterial pressure of 40 to 45 is achieved with systolic pressure in the range of 55 to 65 and a heart rate in the range of 140 to 150 bpm. Management is guided by mixed venous oxygen saturation (SvO_2) measurements from superior vena cava samples. Frequently, SvO_2 is continuously monitored using an oximetric superior vena cava catheter. Adequate systemic oxygen delivery is indicated by maintaining $\text{SVO}_2 > 45\%$ and an arterial venous O_2 difference of < 30 . Excessive vasodilation caused by POB is managed with vasopressin.¹² Pharmacologic paralysis is maintained throughout the first 24 to 48 hours. Delayed sternal closure is used in most patients with closure when tissue edema subsides, usually postoperative day 3 to 5. Placement of a peritoneal drain is part of our routine management.

Data Acquisition and Analysis

The study was approved by the Institutional Review Board of the Hospital for Sick Children. Medical records and the surgical database were reviewed for demographic, operative and perioperative, and outcome data. Diagnosis and morphological characteristics were obtained by echocardiography.

Definitions

Early circulatory collapse was defined as either failure to come off CPB, resulting in extracorporeal membrane oxygenation (ECMO) support or death, or any case of sudden circulatory collapse requiring cardiopulmonary resuscitation and/or institution of ECMO support during the initial 72 hours. Typically, the infant appeared clinically stable and had circulatory collapse for no apparent reason or during minimal stimulation such as suctioning. A technical issue was defined as sudden circulatory collapse in which the cause was attributed to mechanical or technical problems. Coronary ischemia, shunt thrombosis, arch gradient requiring reoperation, and failed biventricular repairs are included in this definition.

Data Analysis

Goals of this analysis include defining the time-related prevalence of and risk factors for early circulatory collapse after the Norwood operation. All analyses were performed using SAS statistical software (version 8; SAS Institute, Inc). Data are described as frequency with percentage, median with range, or mean \pm SD as appropriate.

Time to early circulatory collapse was analyzed. Nonrisk-adjusted nonparametric estimates of time-related freedom from early circulatory collapse were plotted as Kaplan–Meier curves. Modeling the instantaneous risk (or hazard function), searching for multiple phases of hazard, and generation of the characteristic equation of the overall hazard function allowed computation of parametric estimates of freedom from early circulatory collapse.¹³

Risk factors for early circulatory collapse were sought by multivariable analysis of the parametric model with each variable noted in Table 1. Variables associated with ≤ 5 events were eliminated from consideration to minimize model over-determination. Variable selection was by bootstrap bagging; 500 bootstrap data sets of same size as the original data set were analyzed by automated stepwise regression with entry criterion of $P \leq 0.1$ and selection criterion of $P \leq 0.05$. Variables occurring in 50% or more of analyses were entered into the final model and retained when $P \leq 0.05$. Solution of the multivariable model allowed demonstration of the magnitude of effect of selected variables on predicted outcomes.

Results

Prevalence of Early Circulatory Collapse After Norwood Operation

Early circulatory collapse during the first 72 hours occurred in 25 infants (24%). Collapse occurred within 24 hours after

TABLE 1. Variables Examined in Multivariable Analysis of Risk Factors for Early Circulatory Collapse

Demographic
Gender
Age at admission, days
Age at operation, days
Weight at operation, kg
Prenatal diagnosis
Date of operation
Diagnosis and Morphology
Classic vs. variant HLHS
Degree of atrioventricular valve regurgitation
Ascending aortic diameter, mm
Preoperative Clinical Condition
Use of mechanical ventilation
Use of inotropes
Highest lactate level
Intraoperative Data
Cardiopulmonary bypass time
Aortic cross clamp time
Circulatory arrest time
Use of selective cerebral perfusion
Method of arch reconstruction
Shunt size indexed to body weight
Perioperative Management
Use of phenoxybenzamine
Technical issue
Use of ECMO

operation in 11 infants. Freedom from early circulatory collapse was 90% at 6 hours, 84% at 24 hours, 81% at 48 hours, and 79% at 72 hours (Figure 1A). There was a single, rapidly declining early phase of hazard for early circulatory collapse (Figure 1B).

Risk Factors for Early Circulatory Collapse

Three independent risk factors for early circulatory arrest were identified (Table 2). Technical issue at operation and longer cross-clamp time were associated with increased risk, whereas use of POB was associated with decreased risk.

The impact of POB administration for a technically successful operation is shown in Figure 2. Predicted freedom from early circulatory collapse at 72 hours is 95% with the use of POB compared with 69% without ($P \leq 0.002$).

Figure 3 illustrates the impact of technical failure on freedom from arrest during the first 72 hours after the Norwood operation. Technical failure with or without the use of POB was associated with dismal prognosis.

Causes of Early Circulatory Collapse

Twelve events (11%) were explained by technical problems (see technical failures section). Thirteen (12%) patients who appeared clinically stable after the operation had unexpected early circulatory collapse. There was no apparent cause or it

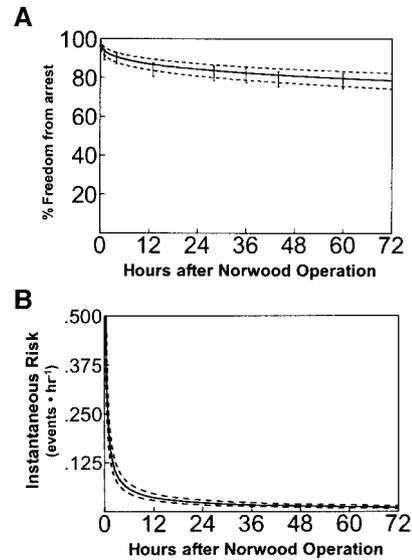


Figure 1. Time-related prevalence of early circulatory collapse. A, Overall freedom from sudden collapse is 90% at 6 hours, 84% at 24 hours, 81% at 48 hours, and 79% at 72 hours. B, Hazard function or instantaneous risk of sudden arrest during the first 72 hours. The risk of arrest is greatest within the first 12 hours after surgery, but the risk does not decrease to zero by 72 hours. Solid lines represent parametric point estimates; dots and error bars represent nonparametric estimates; and dashed lines enclose 90% confidence interval.

was in association with the trauma of suctioning the airway or other subtle event that affect either pulmonary or systemic resistance. Sixteen out of the 25 circulatory collapse infants died.

Outcome of Infants After Early Circulatory Collapse not Related to Technical Failure (n=13)

Six neonates could not be resuscitated and died. Two others were initially successfully resuscitated but died (postoperative day 8 and day 19) from sepsis. Two neonates were placed on ECMO, one survived to hospital discharge and the other died. Four infants survived to hospital discharge, including the one who required ECMO for 36 hours. Table 3 shows the time of early arrest and outcome of infants who had early circulatory collapse unrelated to technical failure.

Outcomes of Infants After Early Circulatory Collapse Related to Technical Failures (n=12)

Twelve neonates had early circulatory collapse that was attributed to technical problems. Eight neonates died, and 4 neonates survived to hospital discharge. Table 4 shows type of technical failure and respective outcomes.

Four neonates had coronary ischemia. Two required ventricular assist device support for acute right ventricular failure

TABLE 2. Risk Factors for Sudden Early Circulatory Collapse

Variable	Direction of Risk	P	Reliability* %
Technical issue at operation	Increase	<0.001	89
Longer cross-clamp time	Increase	0.007	50
Use of phenoxybenzamine	Decrease	0.002	60

*Reported as frequency of selection from 500 bootstrap data sets.

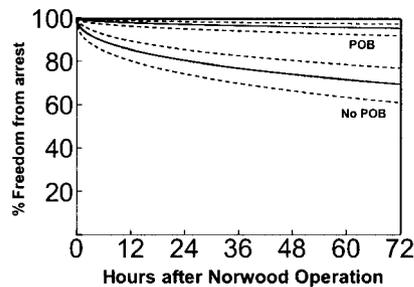


Figure 2. Impact of phenoxybenzamine administration on prevalence of early circulatory collapse. Graph generated by solution of multivariable model with and without phenoxybenzamine (other predictors: no technical failure, cross-clamp time 55 minutes). Solid lines represent predicted parametric point estimates; dashed lines represent 90% confidence interval.

caused by coronary obstruction. Both survived, one after successful surgical revision of the proximal arch anastomosis and one after heart transplantation (after a failed bypass graft to the posterior descending coronary artery and failure to wean from ventricular assist device). Two other neonates died of coronary ischemia; one had a documented right coronary arterial thrombus and infarct on autopsy and one died in the operating room from acute myocardial failure and ischemia.

Two neonates had aortic arch obstruction and underwent successful reoperation after initial circulatory collapse. Two other neonates failed biventricular repair before the Norwood

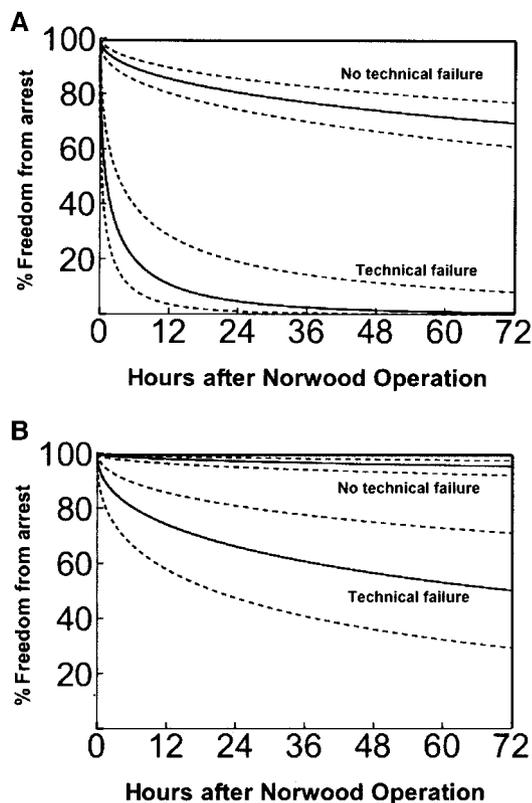


Figure 3. Impact of technical issue on early circulatory arrest within the first 72 hours after operation without (A) and with (B) phenoxybenzamine administration. Graph generated by solution of multivariable model with cross-clamp time 55 minutes. Solid lines represent predicted parametric point estimates; dashed lines represent 90% confidence interval.

TABLE 3. Time After Operation of Sudden Early Circulatory Collapse and Respective Outcomes After Norwood Operation

Infant	Time of Collapse (h)	Outcome
1	1	Death (day 8)
2	1	Death (day 0)
3	1	Death (day 0)
4	3	Discharge
5	4	Death (day 0)
6	8	Death (day 0)
7	8	Discharge
8	10	Discharge
9	10	Death (day 0)
10	13	Death (day 0)
11	28	Death (day 19)
12	36	Discharge
13	60	Discharge

operation. One died of sepsis after an attempted VSD closure/arch reconstruction and a Norwood operation and the other died after an attempted repair of interrupted aortic arch and VSD closure with failure to wean from bypass, ECMO support followed by an attempted Norwood operation, and death. The modified Blalock-Taussig shunt (MBTS) was the source of major complications and death in 4 patients. One died of early shunt thrombosis, 1 required early ECMO and died after revision of MBTS, and 2 others died of sepsis after initial collapse and early revisions of MBTS.

Laboratory Results

There was a trend toward lower intensive care unit admission lactate levels when selective cerebral perfusion was used (7.9 ± 4.9 versus 6.3 ± 3.5 , $P=0.08$). Postoperative lactate levels clearance was not different in patients with and without use of POB (median 6 [0 to 36 hours] versus median 8 [0 to 42] $P=0.3$, respectively). After adjusting for the frequency of technical failures, lower SvO₂ was associated with circulatory collapse (50% versus 37%, 50% versus 39%, and 53%

TABLE 4. Technical Issues and Respective Outcomes After Norwood Operation

Infant	Technical Failure	Outcome
1	Coronary ischemia	RV infarct, death (day 0)
2	Coronary ischemia	Intraoperative death (day 0)
3	Coronary ischemia	Reoperation, discharge
4	Coronary ischemia	ECMO, transplant, discharge
5	Arch obstruction	Reoperation, discharge
6	Arch obstruction	Reoperation, discharge
7	Failed biventricular repair	ECMO, death (day 7)
8	Failed biventricular repair	Sepsis, death (day 25)
9	Shunt occlusion	Death (day 1)
10	Shunt occlusion	ECMO, revision, death (day 13)
11	Shunt occlusion	Revision, sepsis, death (day 48)
12	Shunt occlusion	ECMO, revision, death (day 10)

versus 35% at 4, 12, and 24 hours, respectively) (all $P < 0.005$).

Discussion

Our results indicated that technical issues related to the operation, longer cross-clamp time and no use of POB are independent risk factors for early circulatory collapse after the Norwood operation. Of those, optimal surgical technique is the most important. The arch reconstruction, coronary perfusion, and the MBTS must work in harmony to optimize outcomes in these critically ill infants. Longer cross-clamp time is a reflection of myocardial injury. An added margin of support is obtained with the use of aggressive afterload reduction with POB.

Hemodynamic stability in the early postoperative period after the Norwood palliation is dependent on an adequate cardiac output and a balanced Qp:Qs. The systemic oxygen saturation alone is a poor predictor of Qp:Qs because of variability in SvO₂ and pulmonary vein saturation (SpVO₂) after the Norwood operation¹⁴

Charpie et al reported on the evaluation of systemic perfusion by monitoring the oxygen delivery at tissue level using serum bicarbonate and lactate levels.¹⁵ In his study all negative events were preceded by an abrupt increase in lactate. Furthermore, all patients with good outcome returned to normal lactate level by 24 hours. He concluded that serum lactate may be a useful marker for the need for intervention in the postoperative period.¹⁵ In our study lactate levels were not a sensitive predictor of poor early outcome. The lactate level can be a relatively late sign of systemic hypoperfusion secondary to pulmonary overcirculation.

Rossi et al reported on the importance of monitoring SvO₂ for assessment of cardiac output and systemic perfusion after the Norwood operation.¹⁶ More recently the use of alpha blockade has been introduced with remarkable improvement in survival after the Norwood operation.⁷⁻⁹ The alpha blockade strategy centers on the decrease of the energy requirement and oxygen consumption from the injured postoperative neonatal myocardium. It is well-established that the energy expenditure is greater when the heart is faced with increase afterload than when it is ejecting a larger volume of blood against a lower pressure. During isovolumic contraction, the energy absorbed by shape changes and elasticities in the ventricular walls is much greater when the ventricle develops pressure than when it ejects volume.¹⁷ Simply stated, pressure generation by the myocardium is energetically more costly than volume ejection. On the alpha blockade strategy, manipulation of the myocardial oxygen consumption is obtained by control of heart rate, control of preload, and mainly aggressive afterload reduction.¹⁰ POB promotes irreversible binding of alpha receptors, the serum half-life is ≈ 12 hours, and the effect may last for days because new receptors must be synthesized.

Although the infants are hemodynamically more stable, attention to detail continues to be essential. There is a risk of excessive vasodilatation causing hypotension, with high SvO₂, low AVO₂ difference, tachycardia, and possible signs of myocardial ischemia.¹⁰ Epinephrine infusion may exacerbate the syndrome by promoting further vasodilatation via beta

receptors in the presence of irreversibly blocked alpha receptors. Vasopressin seems to be the ideal drug to overcome the effects of POB under these circumstances because of selective action through a different receptor. The use of vasopressin has improved our ability to manage maximal vasodilatation safely.¹²

This study and others have demonstrated that the use of POB was associated with higher SvO₂ reflecting a higher systemic output when compared with the traditional management. The effect of POB appears to be related to the increase SvO₂ and improved end-organ perfusion.^{8,9}

The identification of excessive pulmonary flow at the expense of systemic blood flow and associated hemodynamic decompensation has led to the use of smaller BT shunts by some groups. In a review of 122 postmortem cases, excessive pulmonary blood flow was the second most common cause of death after coronary ischemia.¹⁸ Others groups have reported that the BT shunt size was a risk factor for death.^{4,19} In our study, the shunt size was not a risk factor for early sudden arrest.

Twedell et al, also using an alpha blockade strategy, did not identify an independent impact of shunt size on SvO₂ and delta AVO₂.⁹ We can speculate that a larger shunt in the presence of alpha blockade strategy is not necessarily a risk factor and may be associated with better oxygen delivery overall.

Kitaichi et al in a canine model of univentricular heart demonstrated a correlation between pulmonary flow and manipulations on PaCO₂ and FiO₂ in a small shunt model.²⁰ The correlation was lost in the larger shunt model. These findings strongly suggest more difficulties in regulating Qp:Qs by the traditional management in larger shunt size neonates. This difficulty may be ameliorated by the use of POB and alpha blockade.

For a patient who is systemically dilated on POB, the Qp:Qs may be insensitive to manipulation in pulmonary vascular resistance because the pulmonary shunt limits pulmonary blood flow mechanically. In other words, after a certain level, an increase in oxygen saturation causing possible increase in pulmonary flow will not cause associated decrease in SvO₂ because of systemic hypoperfusion, because the MBTS act as a fixed resistor limiting the pulmonary flow after a certain level.

Although several morphological risk factors have been defined in the literature, in our study morphological subgroups and the size of the ascending aorta were not risk factors.

Limitations of the Study

Although our technique has continue to evolve and other modifications in our strategy included the use of selective cerebral perfusion, which started at approximately the same period, we believe the use of alpha blockade is the single most important variable in decreasing the incidence of sudden collapse in a neonate who had a favorable technical operation. Modifications in our perioperative care are mirrored by the improved results.

Conclusions

Optimal surgical technique is the most important factor in preventing sudden circulatory collapse after the Norwood operation. An added benefit is obtained with the use of aggressive afterload reduction with POB. The effect of POB appears to be related to increase SvO₂. Lactate levels were not a sensitive predictor of poor early outcome.

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