

unrelated defects may be the result of a common mechanism: altered neural crest cell migration. Further embryonic and genetic experimentation will likely clarify the role of the neural crest cell at the various levels of cardiac development.

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# Novel management strategy for severe cyanosis after Sano modification of the Norwood procedure

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**S**ano modification of the Norwood procedure has resulted in a more favorable distribution of systemic, pulmonary, and coronary blood flow. However, frequently these patients present with severe cyanosis caused by dynamic proximal shunt stenosis. The purpose of this review was to evaluate a management strategy for severe cyanosis in this patient population.

## Methods

Between September 2003 and September 2004, 22 newborns underwent Sano modification at Children's Healthcare of Atlanta. One patient died before hospital discharge and was excluded from further analysis. Four (19%) of the 21 remaining patients presented at  $53 \pm 26$  days of age with oxygen saturations of less than 70%. These 4 patients (group 1) were compared with the remaining 17 (group 2). Management strategy to treat cyanosis in group 1 patients consisted of increasing preload and  $\beta$ -blockade. Age-matched hemodynamic data were evaluated between group 1 before therapy and group 2; variables before and after therapy in group 1 were also evaluated.

## Results

Sano shunt size was 6 mm ( $n = 3$ ; birth weight,  $>3$  kg) and 5 mm ( $n = 1$ ; birth weight,  $<2$  kg; group 1) versus a shunt size of 6 mm ( $n = 13$ ; 4 patients with a birth weight  $<3$  kg; 9 patients with a birth weight  $>3$  kg) and 5 mm ( $n = 4$ ; birth weight,  $<3$  kg; group 2). Hemodynamic variables before and after therapy in group 1 are shown in Table 1. In addition, comparison was made between hemodynamic variables at the time of presentation with cyanosis group 1 and group 2 at the same age (Table 2).

This management strategy prolonged the timing to bidirectional Glenn shunt by  $70 \pm 40$  days in group 1. A bidirectional Glenn shunt was performed at  $123 \pm 32$  days in group 1 (4/4 patients) versus  $133 \pm 28$  days in group 2 (11/13 patients,  $P = .6$ ).

## Discussion

Secondary to the variable pulmonary and systemic vascular resistance ratio, the net effect of the Norwood procedure with a systemic-pulmonary artery shunt is runoff from the systemic to the pulmonary bed during diastole, resulting in a low diastolic blood pressure, a wider pulse pressure, and a change in the pattern of

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**TABLE 1. Hemodynamic variables before and after therapy in group 1**

	Before therapy	After therapy	P value
No. of patients	4	4	1.0
Oxygen saturation (%)	62 ± 9	80 ± 3	.01
Heart rate (beats/min)	151 ± 13	129 ± 9	.03
Respiratory rate (breaths/min)	60 ± 10	50 ± 22	.4
Systolic blood pressure (mm Hg)	90 ± 1	90 ± 12	1.0
Diastolic blood pressure (mm Hg)	44 ± 8	49 ± 13	.5
Mean blood pressure (mm Hg)	60 ± 5	63 ± 11	.5
RVOT proximal gradient (mm Hg)	92 ± 12	58 ± 13	.02

Mean ± SD; paired 2-tailed *t* test. RVOT, Right ventricular outflow tract.

**TABLE 2. Hemodynamic variables at the time of presentation with cyanosis in group 1 and patients in group 2 at the same age**

	Group 1	Group 2	P value
No. of patients	4	17	
Age at the time of the operation (d)	5 ± 3	5 ± 3	1.0
Birth weight (kg)	3.1 ± 0.8	3.2 ± 0.5	1.0
Cardiopulmonary bypass time (min)	135 ± 16	147 ± 28	.3
Total ischemic time (min)	60 ± 3	65 ± 14	.1
Age with cyanosis (d)	53 ± 26	56 ± 13	.8
Oxygen saturation (%)	62 ± 9	80 ± 3	.02
Heart rate (beats/min)	151 ± 13	142 ± 11	.3
Respiratory rate (beats/min)	60 ± 10	52 ± 11	.3
Systolic blood pressure (mm Hg)	90 ± 1	85 ± 8	.5
Diastolic blood pressure (mm Hg)	44 ± 8	48 ± 8	.5
Mean blood pressure (mm Hg)	60 ± 5	60 ± 7	.9
RVOT proximal gradient (mm Hg)	92 ± 12	44 ± 17	.0003

Mean ± SD; paired 2-tailed *t* test. RVOT, Right ventricular outflow tract.

coronary perfusion.<sup>1</sup> Sano modification, substituting a right ventricle–pulmonary artery shunt in place of the systemic–pulmonary artery shunt, provides a more favorable distribution of systemic, pulmonary, and coronary blood flow. Several studies have reported a hemodynamic profile with higher diastolic pressure,<sup>2,3</sup> narrowed pulse pressure,<sup>2,3</sup> and higher coronary perfusion pressure<sup>2</sup> after a right ventricle–pulmonary artery shunt compared with that after a systemic–pulmonary artery shunt.

In our recent experience, 4 (19%) of 21 patients after Sano modification presented at 53 ± 26 days of age with systemic oxygen saturation of less than 70% caused by dynamic proximal shunt stenosis. The proximal shunt Doppler gradient determined by transthoracic echocardiography was 92 ± 12 mm Hg. The gradient was due to dynamic obstruction below the ventricular anastomosis site of the right ventricle–pulmonary artery shunt. We believe that this mechanism of significant cyanosis after the Sano modification is similar to that of hypercyanotic episodes in tetralogy of Fallot: dynamic outflow obstruction. Therefore, we adopted a management strategy of increased preload and  $\beta$ -blockade similar to the treatment strategy for the hypercyanotic episode in patients with tetralogy of Fallot.

Increasing preload was achieved by volume bolus (3 patients) and decreasing diuretic therapy (4 patients).  $\beta$ -Blockade

therapy was with propranolol (1 mg · kg<sup>-1</sup> · d<sup>-1</sup>) in 3 patients and esmolol in 1 patient. This patient was converted to sotalol instead of propranolol because of atrial dysrhythmias not responsive to propranolol. This combination therapy resulted in a 22% increase in systemic oxygen saturation (from 62% ± 9% to 80% ± 3%), with a 15% decrease in heart rate (from 151 ± 13 to 129 ± 9 beats/min).

Increased preload in these single-ventricle patients results in increased systemic resistance, allowing more blood flow into the pulmonary circuit. Increased cardiac output and pulmonary blood flow leads to increased systemic oxygen saturations.  $\beta$ -Blocker therapy has been shown to increase systemic saturation in patients with tetralogy of Fallot through a variety of complex mechanisms, including increases in peripheral resistance,<sup>4</sup> prevention of tachycardia, and shift in the hemoglobin-oxygen dissociation curve.<sup>5</sup> Increased peripheral resistance again allows for increased pulmonary blood flow in this single-ventricle patient. A slower heart rate allows for increased ventricular filling time and thus increased preload. Shifting the hemoglobin-oxygen dissociation curve results in increased availability of oxygen delivery to the tissues. The overall result is improvement in systemic oxygen saturations.

## Conclusion

The medical management strategy of increasing preload and initiating  $\beta$ -blockade therapy is effective in the treatment of severe cyanosis caused by dynamic proximal shunt stenosis after Sano modification of the Norwood procedure.

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