# **Case Report**



## Use of Beta-Blockers in the Post-Operative Period of Sano Modification of the Norwood Procedure with Persistent Cyanosis

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Persistent cyanosis is a frequent complication in the post-operative period of Sano modification of the Norwood procedure. It may be explained by a dynamic proximal shunt stenosis of the synthetic conduit that links the right ventricle to the pulmonary artery, as detected by echocardiographic study. The use of beta-blockers in the post-operative period has been recently described in order to improve the arterial oxygen saturation. In this report, we describe the use of propranolol in two patients undergoing Sano modification of the Norwood procedure, in whom a gradient reduction in the synthetic conduit, increase in the levels of arterial oxygen saturation, decrease in heart rate, and increase in blood pressure were observed, thus resulting in clinical improvement. We conclude that the use of beta-blockers in these cases was beneficial.

## Introduction

Hypoplastic left heart syndrome (HLHS) is characterized by a variety of congenital heart defects that include significant left ventricular underdevelopment, aortic hypoplasia or atresia, and varying degrees of hypoplasia of the ascending aorta, usually accompanied by mitral valve atresia and/or coarctation of the aorta. Without surgical intervention, HLHS is fatal, with a 25% mortality within the first week of life¹. Historically, HLHS has been managed in two ways: with palliative surgeries based on the reconstruction of the ascending aorta (Norwood procedure and its variations), or heart transplantation.

Despite recent advances in surgical techniques for the reconstruction of the ascending aorta, the perioperative morbidity and mortality associated with the first stage still remain very high. Recently, Sano et al<sup>2</sup> described a modification in the first-stage palliation of HLHS using a synthetic polytetrafluoroethylene conduit between the right ventricle and the pulmonary artery (RV-PA conduit) to provide pulmonary blood flow instead of performing a systemic-pulmonary Blalock shunt.

#### **Key words**

Beta-blockers, Norwood procedure, thoracic surgery, cyanosis, adrenergic beta-antagonists; heart defects, congenital.

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Manuscript received July 10, 2006; revised received January 01, 2007; accepted February 26, 2007.

Preliminary results of this novel operation proved to be favorable, but experience is still limited. New therapeutic measures based on anatomical and pathophysiological knowledge have been used to improve the survival of children undergoing the first-stage palliation<sup>2</sup>. A relatively common event that we have seen in these children is the occurrence of significant post-operative cyanosis with a reduction of the arterial oxygen saturation to levels lower than 70%. This reduction is attributed to the dynamic stenosis that occurs in varying degrees in the proximal anastomosis of the RV-PA conduit that can be observed in echocardiographic studies.

In order to minimize the effects of this dynamic stenosis with a reduction of the blood flow to the pulmonary circulation and subsequent reduction in arterial oxygen saturation, we administered a beta-blocker (propranolol) to two neonates undergoing Sano modification of the Norwood procedure.

## **Case Report**

Two patients with HLHS underwent Sano modification of the Norwood procedure on the 3<sup>rd</sup> and 13<sup>th</sup> days of life, respectively. Both progressed with persistent cyanosis in the postoperative period, maintaining an oxygen saturation lower than 70%. The degree of dynamic stenosis in the proximal anastomosis of the RV-PA conduit was diagnosed and estimated using two-dimensional Doppler echocardiography at bedside. On the 7<sup>th</sup> and 9<sup>th</sup> postoperative days, respectively, the patients were treated with enteral beta-blocker (propranolol) at a dose of 1 mg/kg/day, BID, and were clinically followed-up.

At the beginning of the treatment, patient 1 received dobutamine (10 µg/kg/min), milrinone (0.45 µg/kg/min) and adrenaline (0.03 µg/Kg/min), and remained sedated and intubated, and ventilated in the pressure-regulated volume control mode (RR=27 v/min, TV=30 l/min, FiO2=45%, and PEEP=4 cm H<sub>2</sub>O). At the beginning of the treatment, patient 2 received dobutamine (9.5 µg/kg/min) and milrinone (0.8 µg/ kg/min), and remained in non-invasive nasal CPAP ventilation. In the three subsequent days, no significant change was made in the prescription of vasoactive drugs, nor in the ventilatory parameters. Measurements of RV-PA conduit gradient (pre and 72 hours post-therapy), and of the means of systolic, diastolic and mean blood pressure (taken every hour) in the 72 hours pre and post-therapy, as well as of arterial oxygen saturation (obtained from arterial blood gas drawn every six hours), and standard deviation of all parameters are shown in Table 1.

## Results

The patients presented gradients of 42.8 and 42 mmHg (mean of 42.4 mmHg), and 24 and 28mmHg (mean of

Table 1 - RV-PA conduit gradient and hemodynamic variables before and 72 hours after treatment with beta-blockers

	Before treatment				After treatment			
	Case1	Case2	Mean	SD	Case1	Case2	Mean	SD
Gradient between RV and PA (mmHg)	42.8	42	42.4	±0.5	24	28	26	±2.8
Arterial oxygen saturation (%)	63.9	66.1	65	±1.5	72.3	78.1	75.2	±4.1
Heart rate (bpm)	165	153	159	±8.4	142	124.4	133.2	±12.4
Systolic blood pressure (mmHg)	68.1	82.5	75.3	±10.1	77.8	90.3	84	±8.8
Diastolic blood pressure (mmHg)	48.9	56.3	52.6	±5.2	50.7	57.2	53.9	±4.5
Mean blood pressure (mmHg)	56.5	65.7	61.1	±6.5	61	68.9	64.9	±5.5

SD - standard deviation; RV - right ventricle; PA - pulmonary artery.

26mmHg) before treatment and after 72 hours, respectively.

Arterial oxygen saturation was 63.9 and 66.1% (mean of 65%), and 72.3 and 78.1% (mean of 75.2%) before and after treatment, respectively. Heart rate was 165 and 153 bpm (mean of 159 bpm), and 142 and 124.4 bpm (mean of 133.2 bpm) before and after treatment, respectively. Systolic blood pressure was 68.1 and 82.5 mmHg (mean of 75.3 mmHg), and 77.8 and 90.3 mmHg (mean of 84 mmHg) before and after treatment, respectively. Diastolic blood pressure was 48.9 and 56.3 mmHg (mean of 52.6 mmHg), and 50.7 and 57.2 mmHg (mean of 53.9 mmHg) before and after treatment, respectively. Mean blood pressure was 56.5 and 65.7 mmHg (mean of 61.1 mmHg), and 61 and 68.9 mmHg (mean of 64.9 mmHg) before and after treatment, respectively.

## **Discussion**

HLHS is a condition that causes high mortality even after surgical treatment. Its postoperative management should be individualized for the surgical technique used because in the conventional procedure (Norwood operation) the systemic and pulmonary circulations are in series, whereas in the Sano modification of the procedure, they are in parallel. Also, in the Sano modification of the procedure, there may be a dynamic stenosis of the synthetic conduit in the RV-PA anastomosis, thus leading to persistent cyanosis.

Our study is consistent with Simsic et al's recent publication and showed that the use of beta-blockers in the postoperative period of Sano modification of the Norwood procedure resulted in a reduction of the heart rate, increase in saturation, and reduction of the RV-PA conduit gradient. Although our study had not been designed to quantify the cardiac output and peripheral vascular resistance, we can clinically infer that there was an increase in the cardiac output and decrease in peripheral vascular resistance. With the improvement in cardiac performance and decrease in the degree of stenosis in the proximal anastomosis of the RV-PA conduit, an increase in the pulmonary circulation was observed, with a shift of the oxyhemoglobin dissociation curve, thus providing a significant improvement of cyanosis with an increase in the arterial oxygen saturation.

## Conclusion

The therapeutic strategy with the use of propranolol proved to be effective in the treatment of severe cyanosis caused by dynamic stenosis in the proximal anastomosis of the RV-PA conduit after Sano modification of the Norwood procedure.

#### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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