Resistant Hypoxemia in an Infant With a Right Ventricle–to–Pulmonary Artery (Sano) Shunt

Benjamin Pieters, DO,* Troy A. Johnston, MD,† Thomas K. Jones, MD,† Gordon Cohen, MD, PhD,‡ and Christer Jonmarker, MD, PhD*

INFANTS BORN WITH aortic atresia and a hypoplastic left heart are often palliated with a Norwood procedure.[1] Sano et al[2] have developed a modification in which pulmonary blood flow is provided by a right ventricle (RV)–to–pulmonary artery shunt (Fig 1), instead of a subclavian artery–to–pulmonary artery shunt. Immediately postoperatively, the Sano shunt tends to give higher aortic diastolic pressure and more stable hemodynamics than the traditional shunt,[2,3] but shunt obstruction and early or late RV dysfunction can occur.[2-6] This report describes the authors' experience in the intraoperative management of an infant who developed severe cyanosis several months after a modified Norwood procedure.

CASE REPORT

An 11-week-old, 4.5-kg female patient with aortic atresia and a hypoplastic ascending aorta who had undergone a modified Norwood procedure including placement of a 6-mm Sano shunt at 9 days of age was scheduled for urgent cardiac catheterization because of worsening cyanosis.

The patient had a complicated earlier history. At five weeks of age, 1 month after the Norwood operation, she was admitted with electrocardiographic signs of generalized myocardial ischemia. Cardiac catheterization showed stenosis of the proximal aortopulmonary anastomosis, and ischemia was resolved by placing a stent across the stenosis down into the native segment of the ascending aorta. One week after this procedure, the patient developed focal seizures of the left hand and was placed on antiepileptic medications. At the next follow-up visit at 10 weeks of age, 8 days before the current events, she had a pulse oximeter saturation (SpO_2) of 80%, but an echocardiogram showed aortic arch obstruction and a hypertrophic, systemic RV with decreased function, and Sano shunt narrowing. A cardiac catheterization was performed, showing a systolic RV pressure of 120 mmHg with a 50-mmHg gradient across the proximal Sano anastomosis. The major finding, however, was a severe coarctation of the aortic arch with a systolic gradient of 78 mmHg. The coarctation was located in the distal aortic arch beyond the original Norwood anastomosis. RV angiography documented an additional stenosis in the pulmonary artery between the distal Sano anastomosis and the right lung. A 5-mm stent was placed in the aortic coarctation, and, after balloon dilation, there was no residual gradient. The resultant decrease in systemic outflow resistance caused an immediate decrease in SpO_2 (from 75% to 65%), but, over the next hour, the saturation gradually returned to 70% to 80%. The patient was believed to be a candidate for an early bidirectional upper cavopulmonary connection (Glenn procedure) and was admitted for observation. Over the next few days, however, SpO_2 slowly declined to 50% to 60%. An echocardiogram showed RV hypertrophy but good RV function. There was increased flow velocity in both the proximal and distal Sano anastomoses, and right pulmonary artery flow appeared diminished. There was no improvement with administration of oxygen or diuretics, and she was transferred to the intensive care unit and scheduled for a cardiac catheterization with possible stenting of both the Sano shunt and the pulmonary artery stenosis.

On arrival in the catheterization unit, the infant had a peripheral intravenous catheter in place. She was in mild distress, and her skin color was gray-blue with good capillary refill. The SpO_2 was 55% while breathing 2 L/min of nasal oxygen, the respiratory rate was 53/min, heart rate was 116/min, and blood pressure in the right arm was 78/40 mmHg. She was preoxygenated, given intravenous fentanyl, 1 μg/kg, and propofol, 1 mg/kg, and then observed for a few minutes. The patient became calm, and there was a small decrease in blood pressure but no change in heart rate or oxygen saturation. Another dose of fentanyl, 1 μg/kg, was administered, and, 2 minutes later, propofol, 1 mg/kg, was given followed by vecuronium, 0.1 mg/kg. She was intubated with a 3.0 cuffed oral endotracheal tube and ventilated with 0.3% to 1% sevoflurane in oxygen. The ventilator was set at pressure-controlled ventilation with pressures of 17/5 cmH_2O, an inspiratory: expiratory ratio of 1:2, and a rate of 20/min, resulting in tidal volumes of 65 mL. Shortly after intubation, SpO_2 decreased to below 50%. The systolic blood pressure had decreased to 65 mmHg, and the heart rate was 120/min. Phenylephrine, 5 + 5 μg, and 50 mL of lactated Ringer's solution and 40 mL of 5% albumin were given. This resulted in an increase in systolic blood pressure to over 80 mmHg, but there was no improvement in SpO_2. Fluoroscopy showed good endotracheal tube position, and the patient was easy to ventilate and had no airway secretions. Another 2 doses of phenylephrine, 10 μg, were given, increasing the systolic blood pressure to 90 to 110 mmHg, but SpO_2 remained low. There was improvement in SpO_2 when additional 10-mL/kg boluses of lactated Ringer's solution were administered, but the improvement was not dramatic (about 5% increase in SpO_2) and lasted only 5 to 10 minutes. A right radial artery catheter and an additional peripheral intravenous catheter were placed. The cardiologist then proceeded with the examination through a left internal jugular vein approach. Anesthesia was maintained with fentanyl and sevoflurane in 100% oxygen. The patient remained hemodynamically stable with systolic blood pressures of 70 to 100 mmHg but continued to be severely cyanotic (SpO_2 40%-50%, PaO_2 21-29 mmHg, and a right atrial saturation of 19%). There was a further increase in systolic blood pressure to 110 to 120 mmHg and an increase in heart rate to 140 to 150/min during dopamine infusion, 5 to 10 μg/kg/min, but no increase in SpO_2. There was no noted SpO_2 response to epinephrine, 1 μg/kg intravenously, or inhaled nitric oxide, 40 ppm, but, as before, transient improvements were seen with volume administration. The patient had short episodes of ST-segment depression during manipulations of the angiography catheter but was otherwise hemodynamically stable. The first arterial blood gas, obtained about 1 hour after induction, showed a mild respiratory acidosis (pH 7.38, pCO_2 52, base excess +5), which had been corrected when the next blood gas was obtained 1 hour later (pH 7.46, pCO_2 41, base excess +5).

The RV angiogram showed a severe, intramural narrowing of the proximal Sano shunt (Fig 2). Once it was realized that the obstruction might be muscular in nature, the dopamine was discontinued. Because of concerns that the patient would not tolerate the manipulations

*From the *Department of Anesthesiology and Pain Medicine, †Division of Pediatric Cardiology, Department of Pediatrics, and ‡Section of Pediatric Cardiac Surgery, Department of Surgery, Children's Hospital and Regional Medical Center and the University of Washington School of Medicine, Seattle, WA.*

Address reprint requests to Christer Jonmarker, MD, PhD, Department of Anesthesiology and Pain Medicine, Children's Hospital and Regional Medical Center, 4800 Sand Point Way NE, Seattle, WA 98105. E-mail: christer.jonmarker@seattlechildrens.org

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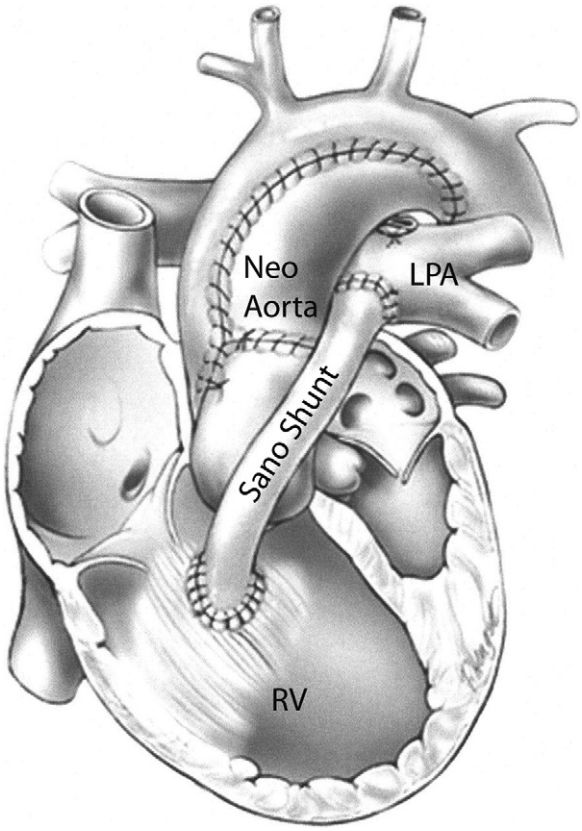

Fig 1. Norwood procedure with Sano shunt. Commonly, a 5- to 6-mm Gore-Tex conduit (W.L. Gore & Assoc, Flagstaff, AZ) is anastomosed to the RV epicardium. LPA, left pulmonary artery. (Modified from Nigro et al,[5] with permission from the Society of Thoracic Surgeons.)

necessary to stent the Sano shunt and because of the foreseeable need for a pulmonary artery dilatation, she was prepared for an emergent "hybrid" Glenn procedure (ie, a combined surgical and catheter intervention) in the catheterization laboratory. A near-infrared spectroscopy monitor probe (INVOS cerebral oximeter; Somanetics, Troy, MI) was placed over the forehead, and the patient was allowed to cool to 36°C. Because the hematocrit had decreased to 30%, secondary to administration of 250 mL of lactated Ringer's and 250 mL of albumin, the patient was also transfused with packed red cells. The operation was performed through a median sternotomy with aortic and bicaval cannulation. The systolic blood pressure remained at 100 to 120 mmHg, but the patient continued to have SpO_2 values of 40% to 50% and near-infrared spectroscopy values of 20% to 25% until normothermic cardiopulmonary bypass was started. After completion of the Glenn procedure, the stenotic segment of the pulmonary artery was stented via a catheter placed through the previous cannulation site in the superior vena cava. The procedure did not require aortic cross-clamping, but the patient's preoperative condition and the relatively long cardiopulmonary bypass time (183 minutes) increased the risk for postoperative bleeding and RV dysfunction. Because of the severity of the proximal Sano obstruction (Fig 2), neither pulmonary overcirculation nor interference with flow through the cavopulmonary anastomosis seemed likely, and the Sano shunt was therefore left in place for later closure. The patient was easily weaned from cardiopulmonary bypass and transferred to the intensive care unit with stable hemodynamics and SpO_2 values of 80%, a PaO_2 of 36 mmHg, and a normal acid-base balance (pH 7.48, pCO_2 35, base excess +3) while ventilated with an F_IO_2 of 0.5. She was extubated 2 days later, transferred to the ward on the fifth postoperative day, and discharged from the hospital on aspirin, diuretics, angiotensin-converting enzyme inhibitors, and antiepileptic medication on the 10th postoperative day. At a follow-up visit in the neurology clinic 2 months later, there were no signs of neurologic injury; she was interactive and developing normally.

DISCUSSION

Although cyanosis in patients with Sano shunts is more commonly caused by a stenosis at the distal shunt anastomosis,[2,3] proximal obstruction has also been reported.[5,6] Nigro et al[5] recently reported sudden death from obstruction of the proximal Sano shunt anastomosis in an infant 3 months after a modified Norwood procedure. In their case, postmortem examination revealed that the obstruction was caused by fibrointimal hyperplasia emanating from the surrounding RV endocardium. Because the Sano shunt is anastomosed to the epicardium, myocardial compression of the intramural shunt segment is also possible and can occur if the subendocardial resection is too limited[2] or, as was the case in the present patient, if RV hypertrophy develops because of increased afterload secondary to recurrent coarctation of the aorta.

In the present patient, the nature and severity of the proximal shunt obstruction were not appreciated during anesthetic induction. At that time, it was believed that the stenosis was fixed and that the recent decrease in saturation was mainly caused by a decrease in Qp/Qs ratio secondary to the aortic angioplasty performed 8 days previously. Under such circumstances, anesthesia induction with ketamine can be a good alternative, but to avoid increasing heart rate and myocardial oxygen consumption, the authors usually prefer to use a combination of fenta-

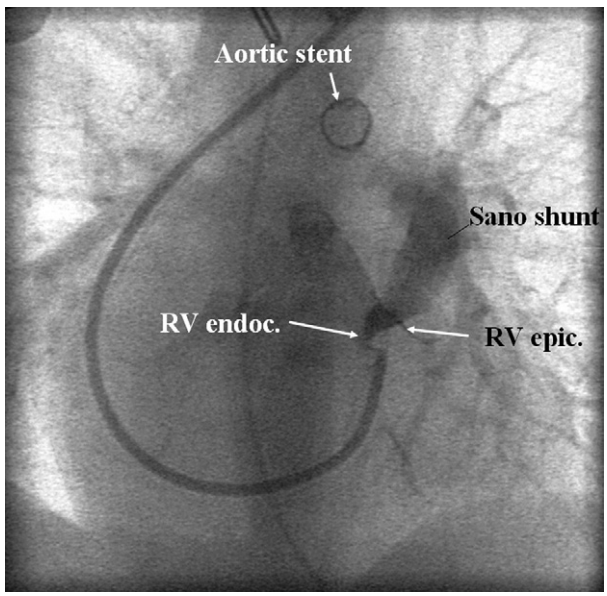

Fig 2. RV angiogram, anterior-posterior view. There is right ventricular hypertrophy and severe stenosis of the inner part of the Sano shunt ventriculotomy. RV endoc., right ventricular endocardium; RV epic., right ventricular epicardium; aortic stent, aortic arch stent placed 8 days earlier (see text).

nyl, propofol, and a vasoconstrictor. Propofol decreases systemic vascular resistance,[7] but hypotension can often be avoided if propofol is combined with small doses of fentanyl and slowly titrated to effect. If the blood pressure should decrease, this can usually be quickly corrected with a small dose of a vasoconstrictor and fluid administration. In the present patient, there was a 10- to 15-mmHg decrease in blood pressure after induction accompanied by a decrease in saturation. Although phenylephrine promptly increased blood pressure to above preinduction values, there was no improvement in oxygen saturation. SpO_2 improved temporarily with volume administration, presumably because this increased RV filling and therefore decreased the obstruction. Neither epinephrine bolus injection nor dopamine infusion improved SpO_2, likely because the Sano stenosis was partially muscular and the increase in RV contractility tended to worsen the obstruction. Dopamine probably also increased myocardial oxygen consumption, and once it became obvious that the RV wall obstruction (Fig 2) was the primary reason for the desaturation, the infusion was discontinued. Instead, administration of β-blockers was considered in order to decrease RV wall tension. Retrospectively, the authors believe it would have been reasonable to try a slow infusion of esmolol, but there was no improvement when deepening the anesthetic with sevoflurane and fentanyl, and at the time of the events, the authors were concerned that the administration of β-blockers would lower the blood pressure and destabilize the situation. While awaiting surgical palliation, the authors therefore focused on maintaining blood pressure, RV filling, optimizing systemic oxygen delivery, and trying to avoid any further increase in heart rate.

The authors believe the slow SpO_2 decrease observed during the week preceding the events was probably an indirect effect of stenting the aortic coarctation; the resultant decrease in afterload improved the function of the hypertrophic RV, thereby worsening the proximal Sano shunt obstruction. Initially, the authors' interpretation of the decrease in SpO_2 after induction of anesthesia was that it was caused by a decrease in systemic vascular resistance secondary to a decrease in endogenous catecholamine release and propofol administration, but the fact that the cyanosis did not improve when systemic vascular resistance was increased suggests that other mechanisms were responsible. Possibly, positive-pressure ventilation may have contributed by decreasing RV filling and increasing pulmonary impedance to shunt flow[8] or by changing the geometry of the Sano shunt or its connections.

In summary, the present patient had a complex cardiac condition with severe hypoxia that worsened with anesthetic induction and did not respond to vasoconstrictors, inotropes, or attempts to lower pulmonary vascular resistance. The principle cause of hypoxia was severe proximal obstruction of the intramural part of the Sano conduit secondary to ventricular hypertrophy. The authors agree with Nigro et al[5] that proximal shunt obstruction should be an early consideration in patients with Sano shunts and worsened cyanosis so that surgical intervention or stenting[6] can be undertaken. The authors' experience also suggests that the distinction between a proximal, possibly dynamic obstruction and a distal, fixed obstruction may be clinically important when hemodynamically managing patients before such interventions.

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