Significance of Low Peak Doppler Velocity in the Proximal Sano Conduit in Hypoplastic Left Heart Syndrome

Shyam K. Sathanandam, MD, Ranjit Philip, MD, Andrew Van Bergen, MD, David A. Roberson, MD, Wei Cui, MD, Michel N. Ilbawi, MD, Alexander J. Javois, MD, and Tarek S. Husayni, MD

The Heart Institute for Children, Advocate Hope Children's Hospital, Oak Lawn, Illinois; and Department of Pediatrics, University of Tennessee Health Science Center, Memphis, Tennessee

Background. **The Sano modification of the Norwood operation is a well-established first step palliation for hypoplastic left heart syndrome (HLHS). Theoretically, the first point of resistance to pulmonary flow should be in the proximal Sano, generating high Doppler flow velocity. Paradoxically, however, some patients have low gradients in the proximal Sano conduit. The objective of this study was to determine the hemodynamic and anatomic significance of low proximal Sano Doppler flow velocity and its clinical implications.**

Methods. **Doppler-derived peak gradients in the proximal Sano conduits were measured in HLHS patients after Norwood-Sano surgery over a 4-year period and confirmed by cardiac catheterization within 2 to 4 weeks. Clinical outcomes of patients with proximal Sano gradients of 30 mm Hg or less (group 1) were compared with patients whose gradient was greater than 30 mm Hg (group 2).**

Results. **Of the 53 patients, 21 (40%) belonged to group 1. Patients in group 1 had smaller ostial right and left pulmonary artery (PA) diameter (3.2 ± 1.2 mm versus 4.5 ± 1.8 mm, $p = 0.03$; and 3.4 ± 1.2 mm versus 5.6 ± 2.1 mm, $p = 0.01$) when compared with patients in group 2. Patients (7 of 10) who needed either balloon angioplasty of a distal Sano or proximal branch PA were from group 1 ($p = 0.01$). Patients in group 1 had higher rates of unintended PA interventions (33% versus 9%) and complications (48% versus 16%) compared with group 2.**

Conclusions. **Low peak Doppler flow velocity in the proximal Sano correlates with the presence of either distal Sano stenosis or proximal branch PA stenosis. These patients require close follow-up in the interstage period and may need earlier intervention.**

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Better understanding of anatomy and physiology of patients with hypoplastic left heart syndrome (HLHS) and advances in surgical techniques and postoperative management have contributed to a remarkable improvement in outcomes after the Norwood operation. The classic Norwood operation involved a modified Blalock-Taussig shunt (mBTS) for pulmonary blood flow. An alternative is the Sano shunt, which involves placing a valveless polytetrafluoroethylene conduit from the right ventricle (RV) to the pulmonary artery (PA) to provide pulmonary blood flow [1].

The advantages of the Sano modification (RV-PA conduit) are associated with the absence of diastolic runoff from the systemic circulation to the pulmonary circulation. Hence, it potentially provides a stable immediate postoperative course, increased coronary arterial flow, improved weight gain related to improved splanchnic perfusion, and lower interstage mortality [2–6]. The concerns of the Sano modification are related to the ventriculotomy, which could contribute to ventricular dysfunction or aneurysm formation, or be a nidus for arrhythmias. Free conduit insufficiency may result in ventricular volume overload and inadequate PA growth, which may be due to the lack of forward flow during diastole [7, 8].

Several case series report improved short-term survival with the Sano modification [1, 3, 9, 10]. Retrospective reviews have demonstrated no early or midterm survival differences between the mBTS or the Sano modification [4, 11–13]. A study using cardiac magnetic resonance showed smaller right pulmonary artery growth and worsening ventricular function in the Sano cohort compared with the mBTS cohort [7]. The Single Ventricle Reconstruction Trial, which is a multicenter, randomized clinical trial, compared the intermediate outcomes of neonates undergoing the mBTS with those of the Norwood-Sano operation [8]. It concluded that neonates who have the Sano modification had smaller PA growth and higher incidence of unintended interventions.

Hemodynamically, the first point of high resistance to pulmonary blood flow is in the proximal Sano conduit

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Address correspondence to Dr Philip, Division of Pediatric Cardiology, Le Bonheur Children's Hospital, Heart Institute, 3rd Flr, 50 N Dunlap St, Memphis, TN 38103; e-mail: rphill15@uthsc.edu.

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Abbreviations and Acronyms

BDG = bidirectional Glenn
HLHS = hypoplastic left heart syndrome
LPA = left pulmonary artery
MA-AA = mitral atresia and aortic atresia
mBTS = modified Blalock-Taussig shunt
MS-AA = mitral stenosis and aortic atresia
MS-AS = mitral stenosis and aortic stenosis
PA = pulmonary artery
RPA = right pulmonary artery
RV = right ventricle
S1P = stage 1 palliation

and should result in a high Doppler flow velocity signal at this point. Paradoxically, however, we observed that a number of patients had low gradients in the proximal Sano conduit. We postulated that a low flow velocity in the proximal Sano conduit is caused by a significant distal obstruction. The purposes of this study were to determine whether the low proximal Sano flow velocity is caused by significant distal obstruction, and to ascertain the hemodynamic, anatomic, and clinical implications of such findings.

Patients and Methods

Review of medical records and computerized hospital data was approved by the Institutional Review Board, and the procedures followed were in accordance with institutional guidelines for retrospective record review and protection of patients' confidentiality. The Institutional Review Board waived the need for patient consent.

Study Population

Neonates with HLHS who were managed at our institution during a 4-year period were included in the study. HLHS was defined as the presence of mitral atresia and aortic atresia (MA-AA), mitral stenosis and aortic atresia (MS-AA), or severe mitral stenosis and aortic stenosis (MS-AS). All had mitral valve diameter, aortic valve size Z –4 or less. All cases with additional or other cardiac anomalies were excluded from this study, including HLHS with ventricular septal defect, unbalanced atrioventricular septal defect, double outlet right ventricle with mitral and aortic stenosis or atresia, critical aortic stenosis, endocardial fibroelastosis with normal or large left ventricle, and heterotaxy syndromes requiring a Norwood operation.

Operative Technique for the Sano Conduit

Our surgical approach for the Sano conduit involves anastomosing the PA conduit at the partially augmented PA wall early during the cooling phase. During the rewarming phase, a modification of the anastomosis to the RV is made, and the conduit directed leftward to the aorta. A full thickness ventriculotomy is performed to construct a transmural opening of 4.5 mm to 5 mm. After the length of the polytetrafluoroethylene conduit is fashioned, its anterior wall is reverse beveled. The posterior wall is then sutured to the epicardial surface of the ventriculotomy. A "baseball-plate" tailored bovine pericardial hood is constructed to augment the anterior anastomotic site. This hood is fashioned purposely redundant. It is believed to reduce the diastolic regurgitant flow due to dynamic anterior wall effect and to prevent muscular hypertrophy, shunt intimal hyperplasia, and early stenosis at the ventricular end of the conduit.

Measurement Variables

We measured Doppler-derived peak gradient in the proximal Sano conduit in HLHS survivors after Norwood-Sano surgery. Pulse-wave Doppler was performed in a high parasternal short-axis view or, rarely, from a subcostal sagittal view of the Sano conduit. These views provide the best pulse Doppler sampling of the proximal Sano conduit. All patients underwent cardiac catheterization within 2 to 4 weeks of the Doppler study before referral for the bidirectional Glenn (BDG) surgery. We confirmed Doppler-measured peak Sano gradients by cardiac catheterization. Patients who had Doppler proximal Sano gradient of 30 mm Hg or less (group 1) were compared with patients who had Sano gradient greater than 30 mm Hg (group 2). Patients were followed till Fontan completion.

The McGoon ratio is the combined diameter of the right pulmonary artery (RPA) and left pulmonary artery (LPA) at the level of the hilum of the lung, to the diameter of the descending thoracic aorta at the level of the diaphragm. We calculated the McGoon ratio using the narrowest PA branch diameter, which is at the level of the ostium of each branch PA and also at the level of the hilum of the lungs. They are called ostial McGoon ratio and hilar McGoon ratio, respectively. The Nakata index is the sum of the cross-sectional areas of the RPA and LPA at the level of the hilum of the lungs, indexed to body surface area. We calculated an ostial Nakata index and a hilar Nakata index. These indexes were calculated in all patients from PA angiograms. Pulmonary angiography was performed by contrast injection in the proximal Sano conduit with the lateral camera in the 90-degree left anterior oblique projection and the anterior-posterior camera in a 45-degree caudal angulation. This view, called the "sleeping-bat view," provides the best layout of the entire Sano conduit and the branch PAs without any foreshortening.

The following were considered unintended interventions before the BDG surgery: balloon angioplasty or stent implantation in the Sano conduit or the branch PA, mBTS before the BDG, or PA arterioplasty during the BDG. Mortality, extracorporeal membrane oxygenation (ECMO) support or cardiac transplantation after the BDG were considered unintended complications after BDG. Similarly, PA intervention before or during Fontan or need for a fenestrated Fontan were considered unintended interventions for the Fontan surgery. After Fontan completion, mortality, ECMO support, cardiac