Modified Norwood Procedure for Hypoplastic Left Heart Syndrome

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Background. Since June 1994, we have used a modification of the Norwood technique in 13 patients presenting with the hypoplastic left heart syndrome or similar variants.

Methods. This technique involves coarctation repair, arch reconstruction, and creation of a neo-ascending aorta using autologous great vessel tissue only. Pulmonary blood flow is provided by a central shunt of 3.0- to 4.0-mm Gore-Tex.

Results. The mean age and weight at operation were 15 days (range, 1 to 77 days) and 3.2 kg (range, 1.7 to 4.6 kg), respectively. The mean circulatory arrest time was 32 minutes (range, 25 to 50 minutes). There was one operative death, and there have been no late deaths. Seven patients have gone on to conversion to a bidirectional cavopulmonary shunt at a mean age of 6 months. There have been no cases of recurrent coarctation, arch obstruction, or left pulmonary artery stenosis. Significant coronary insufficiency requiring revision of the ascending aortic reconstruction has developed in 2 patients.

Conclusions. We believe this approach offers the advantage of using the patient's own native tissue for all great vessel reconstruction. This technique may also allow 10 to 15 minutes less of circulatory arrest time. The theoretic benefits include improved growth of repaired structures and avoidance of homograft or prosthetic material. The long-term results remain to be elucidated.

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A staged operative approach is now a well-accepted management strategy in patients born with the hypoplastic left heart syndrome (HLHS) or similar variants. This approach employs an initial palliative procedure designed to align the systemic right ventricle with the ascending aorta and provide appropriately controlled pulmonary blood flow. In suitable patients, subsequent stages include conversion to a bidirectional cavopulmonary shunt or hemi-Fontan and, ultimately, a completed Fontan.

Norwood and colleagues were the first to describe this novel method for treating what had formerly been a uniformly lethal problem [1–3]. The "Norwood procedure," as it came to be known, used a direct anastomosis between the transected main pulmonary artery and arch and ascending aorta to establish right ventricle-to-aortic continuity. Pulmonary blood flow is provided by means of a central aortopulmonary shunt of polytetrafluoroethylene (Gore-Tex; W.L. Gore & Assoc, Flagstaff, AZ).

Although the original Norwood operation clearly revolutionized the management of HLHS, several technical problems emerged. These include proximal left pulmonary artery stenosis and recurrent aortic coarctation. Because of these findings, the original method was abandoned in favor of using prosthetic material, initially Gore-Tex and now most commonly pulmonary homograft, to augment the ascending aorta, arch, and proximal descending thoracic aorta and establish continuity with these structures and the proximal main pulmonary artery.

The results with this strategy continue to be encouraging. Indeed, some centers are now reporting a greater than 80% survivorship in patients undergoing the first-stage operation [4]. In our minds, however, there remain significant problems with this approach including the long-term fate of the homograft tissue required for the aortic reconstruction and the substantial circulatory arrest time necessary to perform the operation. These considerations have led us to examine a new technique based on modifications of the original Norwood procedure in the treatment of these difficult patients.

Material and Methods

Since June 1994, 13 neonates and infants presenting to the Cleveland Clinic Foundation for management of HLHS or similar variants have undergone a modified Norwood operation (described below) as first-stage palliation of their congenital heart defect. Of these 13 babies, 6 had aortic and mitral atresia with minuscule ascending aortas (< 2.5 mm). Six other babies had varying degrees of hypoplasia of the mitral valve and left ventricle, and aortic valve atresia. All patients were considered to have some degree of aortic coarctation.

The mean age and weight at operation were 15 days (range, 1 to 77 days) and 3.2 kg (range, 1.7 to 4.6 kg), respectively. Ten patients were "stable" in the intensive care unit with ductal patency being maintained via prostaglandin E_1 infusions. One patient was transferred in a poor hemodynamic state after attempted percutaneous balloon valvotomy of a severely dysmorphic, hypoplastic

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Fig 1. Schematic representation of a "typical" hypoplastic left heart syndrome patient with diminutive ascending aorta, hypoplastic arch, and aortic coarctation.

Fig 2. After ductal and right atrial cannulation, the proximal ductus is ligated. The main pulmonary artery is transected obliquely between right and left pulmonary arteries with care to avoid the orifices. The bifurcation is patch-augmented with autologous pericardium.

unicusped aortic valve with a 4.0-mm annulus. Two patients presented late, 1 with an unbalanced atrioventricular septal defect at 75 days of life and 1 with HLHS and a patent ductus at 77 days of life.

The technique employed was relatively uniform throughout. After median sternotomy and subtotal thymectomy, a pericardial patch was harvested for later use in reconstruction of the pulmonary artery bifurcation. Cardiopulmonary bypass was established using arterial cannulation of either the ductus arteriosus (10 patients) or ascending aorta (3 patients) and a single right atrial cannula. Pump primes were customized to individual patients using fresh, heparinized whole blood. Our perfusion strategy includes profound systemic vasodilation using phenoxybenzamine (0.25 mg/kg intravenously) and high flow (150 to 200 $mL \cdot kg^{-1} \cdot min^{-1}$) while patients are cooled to a nasopharyngeal temperature of 18°C.

After the institution of cardiopulmonary bypass, the ductus arteriosus is either snared or ligated proximally to limit pulmonary blood flow. The ascending aorta, arch, brachiocephalic vessels, and proximal descending thoracic aorta are widely mobilized. The main pulmonary artery is then transected obliquely starting just proximal to the right pulmonary artery orifice (Figs 1, 2). The untreated pericardial patch is used to augment the pulmonary artery bifurcation. If time permits, a central shunt of 3.0- to 4.0-mm Gore-Tex is created between the innominate artery and proximal right pulmonary artery.

Under deep hypothermia, circulatory arrest is initiated. The heart is protected with a single dose of crystalloid cardioplegia infused via the arterial cannula. The cannula is removed, and the descending aorta is mobilized as distally as possible. A vascular clamp is applied to the distal descending aorta to provide traction and aid in mobilization. All ductal tissue is carefully resected (Fig 3).

The lesser curve of the aortic arch is then opened from left subclavian artery to a point just opposite the innominate artery. The descending aorta is then anastomosed to approximately 50% of the orifice in the arch (Fig 4). The proximal main pulmonary artery is then anastomosed to this composite of descending aorta and arch (Fig 5). A wide atrial septectomy is performed via the right atrial pursestring, and then cardiopulmonary bypass is reinstituted and rewarming begun. While the heart is recovering and the patient is rewarming, the aortopulmonary shunt is completed.

Results

All 13 patients in this preliminary series have undergone a procedure as described to provide first-stage palliation. There has been one hospital death and no late deaths. There have been no early neurologic complications. One patient suffered a mild embolic stroke at 4 months of life, presumably related to an intravenous infusion for treatment of a wound infection. His transient neurologic impairment has resolved.

The mean period of circulatory arrest in this group of

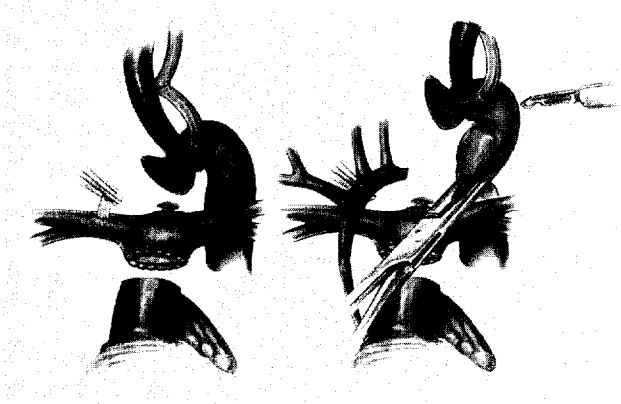

Fig 3. After initiation of circulatory arrest, the descending aorta is clamped. This provides traction for improved mobilization. All ductal tissue is then excised.

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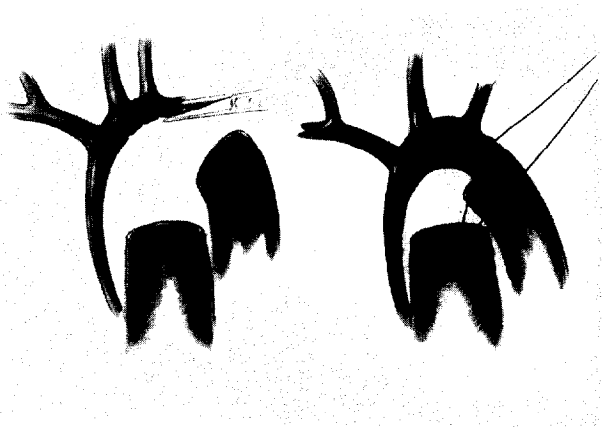

Fig 4. The lesser curve of the aortic arch is opened from left subclavian artery to a point just opposite the innominate orifice. Arch reconstruction is begun by anastomosing the descending aorta to the undersurface of the arch for a distance of approximately 50% of the orifice. The proximal main pulmonary artery is then anastomosed to this composite.

patients was 32 minutes (range, 25 to 50 minutes). The mean period of postoperative ventilation in survivors was 5.5 days (range, 3 to 9 days). The mean duration of intensive care and hospital stay was 8.0 days (range, 5 to 11 days) and 15 days (range, 10 to 21 days), respectively.

Seven patients have undergone cardiac catheterization at ages ranging from 2 to 4 months. There have been no cases of recurrent aortic coarctation. Three patients were thought to have mild proximal right pulmonary artery stenosis. No left pulmonary artery stenosis has developed. All 7 patients in this group have gone on to successful bidirectional cavopulmonary shunt and are well. The remainder of the patients in the overall group are awaiting catheter study.

One important technical problem has occurred in 2 patients and possibly also the patient who died. This has related to patients with long ascending aortas in whom the inferior traction on the arch has resulted in what appeared to be myocardial ischemia due to kinking of the ascending aorta. In 2 patients, the ascending aorta was translocated to the innominate artery with complete resolution of myocardial ischemia. Both patients are alive and well.

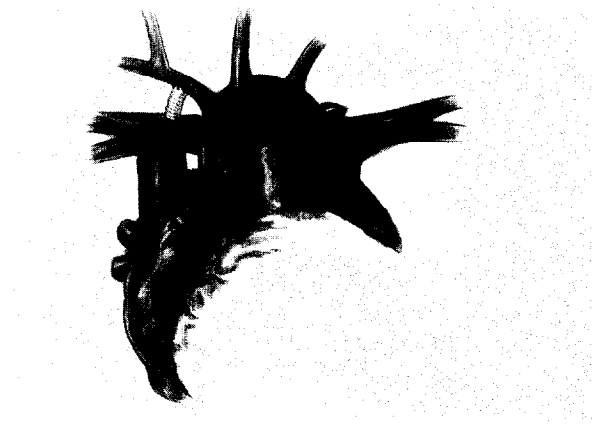

Fig 5. Completed result with aortic reconstruction using autologous great vessel tissue only. A small Gore-Tex shunt (3.0 to 4.0 mm) provides pulmonary blood flow.

As noted, we believe the one operative death was a result of coronary insufficiency. The early postoperative course in this patient with aortic/mitral atresia had been unremarkable. In fact, the patient was successfully extubated on postoperative day 2 with good hemodynamics. Unfortunately, he suffered a sudden cardiorespiratory arrest approximately 24 hours after extubation. Although we were able to adequately oxygenate and ventilate the patient during resuscitation, we could not bring him out of electromechanical dissociation. This had led us to conclude that the shunt was patent but that he likely had myocardial ischemia. A request for postmortem evaluation was denied.

Comment

This report relates a limited experience in a small group of patients. Our intent is to describe what we believe may represent a potential technical advance in the management of these babies. Nonetheless, these results are too preliminary to be able to draw profound conclusions. Clearly, only time and increased numbers of patients will provide the answer of whether this approach measures up to or surpasses the improving results with the "standard" Norwood technique.

We are encouraged, however, by some trends in our data. First, it appears that the early postoperative survival may compare favorably with that of other methods employed in palliating patients with HLHS. As noted previously, others are reporting a greater than 80% survivorship for stage I palliation for HLHS [4, 5]. Although our numbers are entirely too small for a meaningful statistical analysis, the trend is encouraging.

We are also encouraged by the lack of recurrent coarctation or left pulmonary artery stenosis in our series. With regard to the former, we believe an aggressive approach to mobilization of the arch and descending thoracic aorta along with wide resection of all ductal tissues will reduce the tendency for recurrent coarctation. In considering left pulmonary artery stenosis, we believe our technique is an improvement over the original Norwood operation. In that method, the proximal main pulmonary artery was "pulled down" so that it could be anastomosed through the coarctation site. We believe this resulted in an obligatory compression of the left pulmonary artery. The present method "pulls up" the main pulmonary artery to the composite of arch and ascending aorta without impinging on the left pulmonary artery. We also believe the liberal use of autologous pericardium to patch the bifurcation preserves the potential for growth as compared with homograft or prosthetic material.

We are most enthused about what appears to be an

opportunity to offer significantly decreased periods of circulatory arrest. This observation probably rests in a technically more straightforward procedure with shorter suture lines. Again, while not amenable to statistical comparisons, our mean circulatory arrest time of 32 minutes compares very favorably with the mean time of 50 minutes reported in Iannettoni and associates' review [4]. With the ever-increasing knowledge regarding circulatory arrest, all cardiac surgeons are being reminded that previously held ideas concerning "safe" periods of cerebral ischemia may have to be revised. We contend that a savings of 10 to 15 minutes of circulatory arrest time may prove to be the most significant advantage of this technique.

Overall, we are encouraged by our initial results but are not prepared to make long-term pronouncements about this method. The final analysis will require significantly greater numbers of patients and follow-up time.

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