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Original Article

Results of the Norwood operation for hypoplastic left heart syndrome

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EFFORTS TO TREAT BABIES WITH HYPOPLASTIC LEFT heart syndrome began over 30 years ago, when surgeons first reported limited success with palliative surgery. Cayler et al.[1] performed an anastomosis between the ascending aorta and the right pulmonary artery, bilateral banding of the pulmonary arteries, and an atrial septectomy. Their follow-up extended for only 7 months, but the authors predicted that the rapid advances being made in cardiac surgery at the time heralded a new era where hypoplastic left heart syndrome would be cured. Unfortunately, that prediction remains far from true today, although dramatic advances have been made in the therapy of this otherwise lethal condition.

The Norwood operation has remained the mainstay for the initial surgical palliation of hypoplastic left heart syndrome. In this procedure, it is essential to establish an anastomosis between the proximal pulmonary trunk and the augmented aorta so as to provide unobstructed flow between the right ventricle and the aorta, including the coronary arteries. The atrial septum is removed, and the flow of blood to the lungs is regulated by the interposition of a prosthetic tube between the systemic and pulmonary circulations. Despite improvements in outcome, hypoplastic left heart syndrome is still considered "different" from other forms of complex lesions producing the functionally univentricular circulation. The reasons are multiple, but include the technical complexity of the first stage, the reliance on a morphologically right ventricle to support the circulation, interstage mortality, and a past history of poor results. Many issues are of importance in the understanding of proper surgical care, including the use of regional cerebral perfusion, methods of reconstruction of the aortic arch, construction of the appropriate shunt to provide blood for the lungs, and the optimal surgical design of the cavopulmonary connection. This review will focus on the first stage of surgical reconstruction, namely the Norwood procedure itself.

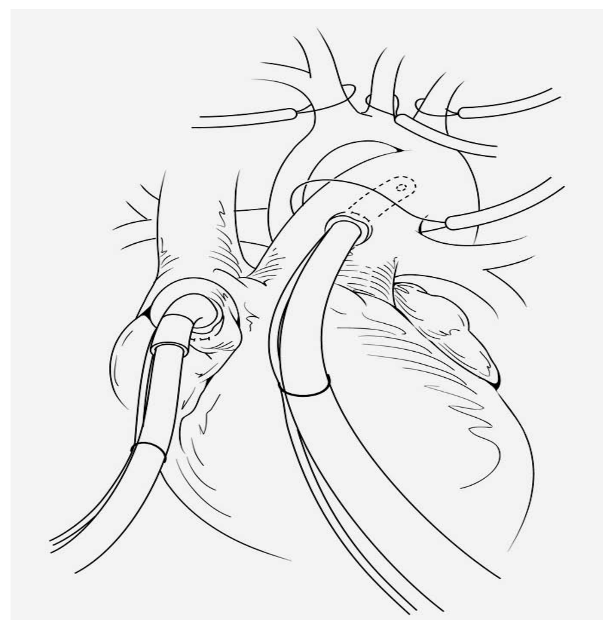

Figure 1.
Cannulation for the Norwood procedure. Snares are placed around the arterial ducts and the brachiocephalic vessels.

Technique

Cannulation for the Norwood procedure is accomplished through the arterial duct and the right atrial appendage (Fig. 1). The atrial cannula is placed more

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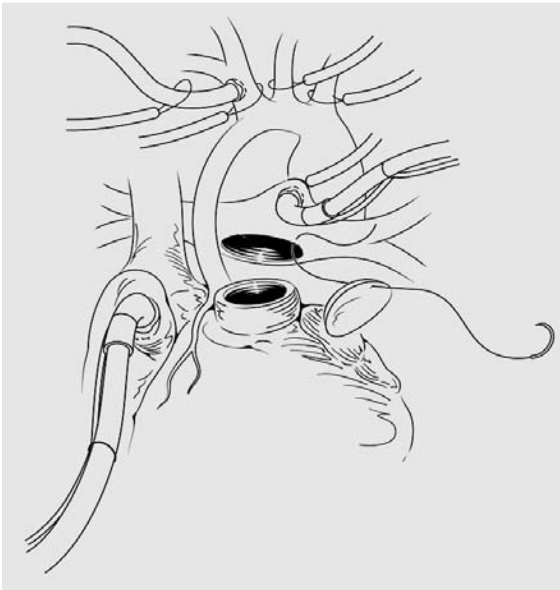

Figure 2.
The pulmonary trunk is divided and the distal portion closed with a patch during systemic cooling. The shunt is placed on the brachiocephalic artery at this time if regional cerebral perfusion is utilized.

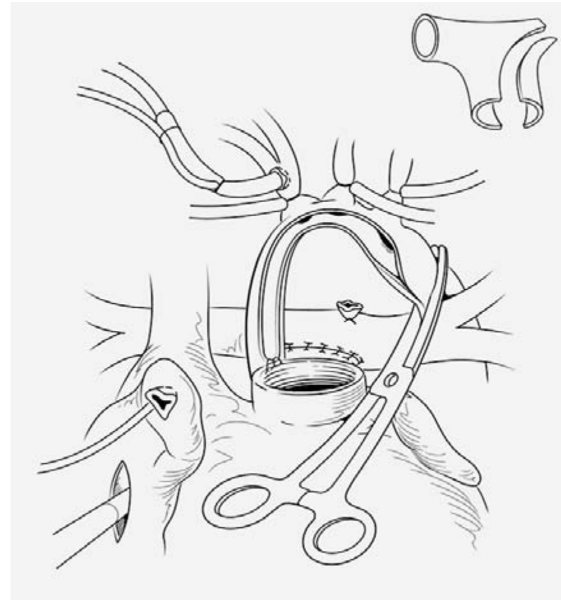

Figure 3.
The duct is excised and the arch opened. The proximal ascending aorta is attached to the adjacent pulmonary trunk. A gusset of pulmonary homograft is trimmed for arch augmentation (upper right). Shown is the position of the cannula for regional cerebral perfusion, and the sucker positioned through the right atriotomy used to excise the atrial septum.

inferiorly on the appendage to preserve as much atrial tissue as possible for the second stage operation, the so-called hemi-Fontan procedure. A snare is engaged around the arterial cannula to exclude the pulmonary circulation during the cooling. The pulmonary trunk is divided just proximal to its bifurcation, and the distal end closed with a patch of Gore-Tex material (Fig. 2). If regional cerebral perfusion is selected, the proximal anastomosis of the shunt, generally a 3.5 or 4-millimetre Gore-Tex conduit, is sutured to the brachiocephalic artery, and the arterial cannula transferred to the shunt to perform selective cerebral perfusion at 25 to 30 cubic centimetres per kilogram per minute into the right carotid artery during reconstruction of the arch (Fig. 3). After cardioplegia is administered through a side arm on the arterial cannula, the snares are adjusted to allow either regional cerebral perfusion or circulatory arrest. The atrial septum is excised and the duct is divided. All ductal tissue is excised, and the aorta opened from the descending aorta beyond the point of ductal insertion to the ascending portion, stopping at the level of the proximal pulmonary trunk. The pulmonary trunk is connected to the adjacent ascending aorta with a few interrupted sutures to establish proper orientation, and prevent any obstruction to coronary arterial flow. Augmentation of the arch is accomplished with the use of a gusset of pulmonary arterial allograft tissue. The distal anastomosis of the shunt is then completed to the pulmonary trunk during full body perfusion and systemic rewarming (Fig. 4).

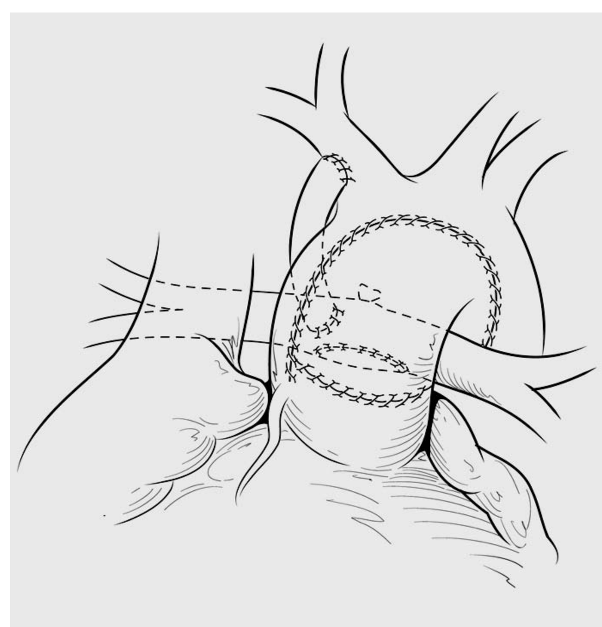

Figure 4.
The completed operation is shown. The shunt is placed between the brachiocephalic and central pulmonary arteries.

Postoperative management: the dilemma of the shunt

The main goal in the postoperative care of the patient following the Norwood procedure is the optimization

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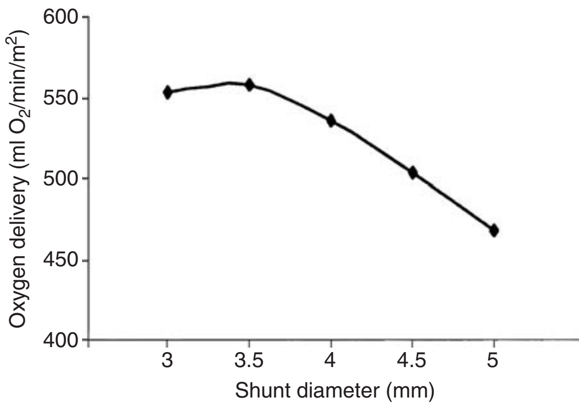

Figure 5.
The relationship between the diamter of the shunt and systemic delivery of oxygen is demonstrated in this graph. Larger diameters result in decreased delivery of oxygen. Reprinted with permission from Migliavacca et al.[4]

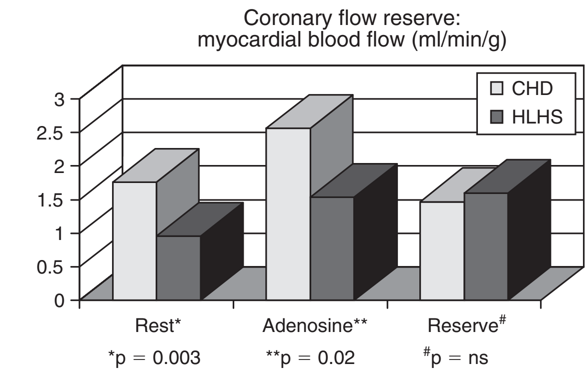

Figure 6.
Rest and maximal coronary arterial flow measured with positron emission tomography in infants following open heart surgery. Controls were compared to patients following the Norwood procedure. Reprinted with permission from Donnelly et al.[6] *CHD: coronary heart disease; HLHS: hypoplastic left heart syndrome.*

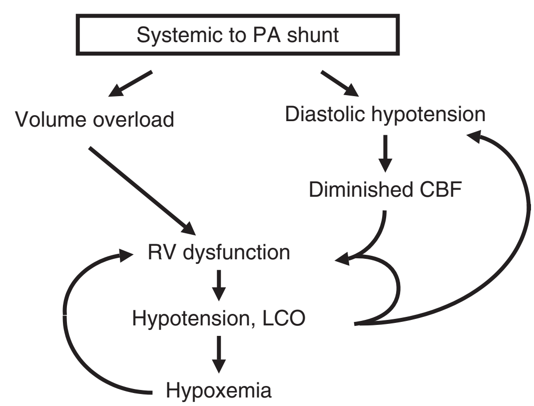

Figure 7.
The vicious circle resulting from a systemic-to-pulmonary arterial (PA) shunt is illustrated. See text for details. CBF: coronary blood flow; RV: right ventricle; LCO: low cardiac output.

of delivery of oxygen to the tissues. Efforts to adjust the resistances of the pulmonary and systemic circulations to achieve the appropriate balance of pulmonary and systemic flows have been the subject of numerous clinical and experimental efforts.[2,3] Data from computer flow modelling has demonstrated that increasing the size of the shunt will result in higher systemic arterial oxygen saturation, but not in higher mixed venous oxygen saturation (Fig. 5).[4] This data has been borne out in clinical experience, where larger shunts have been found to be deleterious, causing an excessive amount of flow of blood to the lungs, even at the expense of systemic flow and oxygen delivery. A ratio of pulmonary to systemic flows of approximately 1 has been shown to be optimal. It should be remembered, nonetheless, that even at a ratio of unity, the right ventricle remains volume overloaded. Systemic vasodilators are important in reducing systemic vascular resistance to optimize this ratio.[5]

It has long been understood that the "Achilles Heel" of the Norwood procedure is the shunt itself. The systemic and pulmonary circulations are placed in parallel, supported by the right ventricle. The most significant difference between this arrangement, and that found in the preoperative state, is the diastolic runoff imposed by the systemic-to-pulmonary arterial shunt. This will result in lower diastolic blood pressure and, potentially, decreased coronary arterial flow. Although little is known about coronary arterial flow in this condition, studies have suggested that resting flow is decreased after the Norwood procedure (Fig. 6).[6] This "competitive" pattern of diastolic flow results in a situation with volume overload, low diastolic pressure, increased right ventricular wall tension, and diminished coronary arterial flow, all of which may serve acutely to destabilize the circulation. A vicious circle results, which often leads to rapid deterioration and death (Fig. 7). Recently, early reports have suggested that a shunt placed between the right ventricle and the pulmonary arteries is associated with more stable hemodynamics and improved early survival following the Norwood procedure. This needs to be balanced against possible right ventricular injury from the incision, increased disturbances of rhythm, and, possibly, a higher prevalence of early failure or occlusion of the shunt.

Outcomes: current results for the Norwood procedure

The mortality following the Norwood procedure has remained high, although a number of centers have reported improvement. In an earlier report from the

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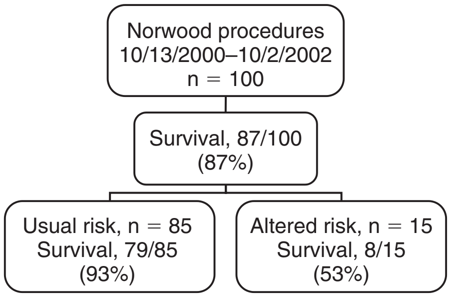

Figure 8.
The results of the most recent 100 consecutive Norwood operations for classic hypoplastic left heart syndrome performed at the University of Michigan.

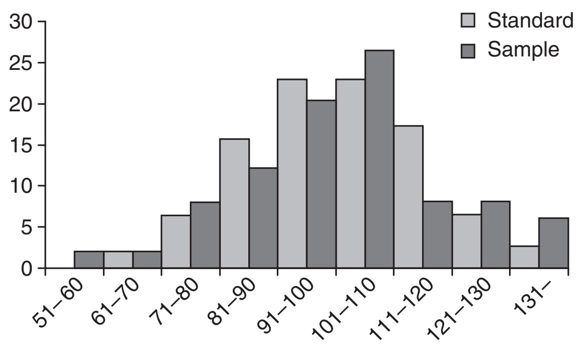

Figure 9.
Combined verbal and motor Wechsler test scores for 50 patients following the Fontan operation. Reprinted with permission from Goldberg et al.[8]

University of Michigan, we reviewed our experience with 158 consecutive patients undergoing surgery between 1990 and 1995.[7] Analysis of risk factors demonstrated that low birth weight or prematurity, significant pulmonary venous obstruction, associated congenital anomalies, and age beyond 1 month were all associated with a significant increase in hospital mortality. Among 31 patients with one or more of these conditions, deemed the group with altered risk, hospital survival was 42% (70% confidence limits between 33 and 51%). Hospital survival was significantly better among the 127 patients without associated conditions, deemed the group at standard-risk, which was 86% (70% confidence limits from 82 to 89%. Importantly, neither the specific anatomy, nor the size of the ascending aorta, emerged as risk factors in that study. Among the most recent 100 consecutive Norwood procedures performed at C.S. Mott Children's Hospital of the University of Michigan between October 2000 and October 2002, hospital survival was 93% for the 85 patients thought to be at standard-risk, and 53% for the 15 patients in the group with altered or high-risk (Fig. 8).

In another study reported from our group, neurodevelopmental outcomes were reviewed in a group of 50 patients, 26 having hypoplastic left heart syndrome, after the Fontan procedures.[8] Combined verbal and motor skills as assessed by standard Wechsler test scores were within the normal range for the entire group, although patients with hypoplastic left heart syndrome scored, on average, less than those with other forms of functionally single ventricle (Fig. 9).

Conclusions

Survival following the Norwood procedure is improving, and recent developments suggest a continued trend in both the quantity and quality of survivors. The major risk remains associated with the first stage procedure, as the risk for subsequent operations remains low.[9,10] The systemic-to-pulmonary arterial shunt as traditionally performed in the Norwood procure has certain immutable hemodynamic disadvantages. These may be overcome with the use of a shunt placed between the right ventricle and the pulmonary arteries, although the potential adverse consequences of a right ventriculotomy must also be considered. There are certain patients, nonetheless, that would appear to benefit significantly by having the shunt constructed between the right ventricle and the pulmonary arteries, such as those with low birth weight and/or prematurity, significant volume overload from tricuspid regurgitation, and possibly those with extremely diminutive ascending aortas, where low diastolic blood pressure might be expected to have its most significant consequences.

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