Aortic reconstruction in hypoplastic left heart syndrome— A reappraisal

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Hypoplastic left heart syndrome (HLHS) is the most frequently encountered of the various congenital heart malformations characterized by a single well-developed ventricle. The introduction more than three decades ago by Fontan and Kreutzer of operative strategies to separate the systemic and pulmonary circulations in patients with tricuspid atresia led to the recognition that all single-ventricle anomalies could be managed in a similar fashion, at least in theory. The unique challenge of aortic atresia–HLHS was that the presence of systemic outflow obstruction and thus ductal dependency of the systemic circulation resulted in rapid and predictable hemodynamic deterioration of neonates with this condition and demanded an operative strategy considerably more ambitious than merely regulating pulmonary blood flow by means of either a pulmonary artery band or a systemic-pulmonary shunt. The availability in the 1970s of prostaglandins to ensure patency of the ductus arteriosus was of major importance in the overall management of neonates with HLHS.

During the same era, numerous investigators and surgical teams experimented with a host of innovative and imaginative strategies to stabilize the circulation of neonates with aortic atresia and related malformations. In addition to the magnitude of the overall challenge, two things became apparent: (1) separation of the systemic and pulmonary circulations by atriopulmonary or cavopulmonary connection could not be achieved successfully in young infants because of the prohibitively high pulmonary vascular resistance and (2) palliative operative strategies that did not preserve the architecture and integrity of the pulmonary arteries and their distal vascular bed only postponed by months the inevitable death that previously had typically occurred during the first few weeks of life.

From these early lessons, Norwood distilled the essential features of successful neonatal palliation of aortic atresia and other forms of HLHS: (1) association of the aorta directly with the right ventricle in a fashion that guarantees unobstructed flow from the right ventricle to the systemic circulation and growth potential for the aorta, obviating the need for further aortic surgery; (2) regulation of pulmonary blood flow to ensure proper growth and development and maturation of the pulmonary vasculature, to avoid the development of pulmonary vascular obstructive disease, and to minimize the volume load on the ventricle; and (3) establishment of a large interatrial communication to avoid pulmonary venous hypertension. Thus, in 1983 Norwood and his associates¹ described successful physiologic repair of aortic atresia–HLHS by means of an initial palliation based on these principles, followed by a modified Fontan procedure.

Physiologic repair of HLHS has evolved from a 2-staged operative strategy, which 10 to 15 years ago was performed in a handful of centers in this country, to a 3-staged operative strategy that is now performed at most major pediatric cardiac surgery centers, not only in the United States but in most parts of the world. The immense value of interposing a second-stage procedure, a hemi-Fontan or bidirectional superior cavopulmonary anastomosis, rapidly became apparent. Limiting the duration of the fragile palliated state to a few months, combined with early reduction of the volume load of the single ventricle, has enabled many more children to eventually undergo successful total cavopulmonary connection. Some unresolved issues remain regarding the optimal timing and exact technical features of the

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eventual total cavopulmonary connection. Nonetheless, it is the initial palliative stage of surgical reconstruction, and the ensuing months characterized by parallel systemic and pulmonary circulations, that still pose the greatest challenges in the overall management scheme.

In this issue of the *Journal*, Poirier and associates² summarize the excellent results of their surgical efforts with a challenging group of neonates and infants with HLHS (80%) and other lesions with ductal dependency of systemic blood flow (20%). As their methods produced an operative survival of 83%, those methods merit careful study. The principal modification of the stage I Norwood procedure that Dr Mee and his surgical team at the Cleveland Clinic espouse is the use of only autologous tissue to reconstruct the aortic arch. This differs from what the authors describe as the “standard Norwood operation,” which includes the use of homograft vascular tissue to augment the amalgamation of the native ascending aorta, the arch, and the proximal main pulmonary trunk. In fact, the procedure described in 1983 by Norwood’s group used only autologous tissue to accomplish this amalgamation.

Over the ensuing 17 years, a variety of materials have been incorporated into the reconstruction, with homograft vascular tissue proving to be the best choice on the basis of availability, handling characteristics, and hemostatic properties. Different surgeons may invoke different rationales for the use of vascular patch material in the stage I palliation. However, more than anything else its use as described by Norwood allows for a standardized operative approach to the neonate with single-ventricle physiology and systemic ventricular outflow obstruction, regardless of specific anatomic details and regardless of the degree of hypoplasia of the ascending aorta and the various elements of the arch. As a result, many of Norwood’s pupils and protégés have come to refer to the stage I palliative procedure as the “omni” procedure, because of its universal applicability. In this operation, the native aorta is incised along its lesser curvature from a point well distal to ductal entry into the thoracic aorta, along the lesser curve of the aortic arch, and down the medial aspect of the diminutive ascending aorta to a point directly opposite the transected pulmonary trunk. The entire arch is augmented with a shield-shaped gusset of cryopreserved pulmonary artery homograft tissue. Proximally, the ascending aorta is anastomosed in side-to-side fashion to the proximal pulmonary trunk, and the amalgamation is completed by suturing the homograft gusset to the remainder of the circumference of the pulmonary trunk. The anastomosis of the ascending aorta to the pulmonary trunk is technically the most critical part. When performed correctly, it leaves a very short segment of native ascending aorta to serve as the immediate pre-coronary arterial pathway. Repeated experience with fashioning the homograft gusset results in an

aortic reconstruction that, while larger than normal proximally, looks very much like “an aortic arch.”

As Norwood did originally, Mee and his associates have chosen to perform the entire reconstruction with autologous tissue. They infer that this might optimize aortic growth and avoid degeneration of patch material. Thus, their modification of the Norwood procedure consists of restructuring the arch by means of a combined anastomosis of the proximal main pulmonary artery to the arch and descending aorta without the use of foreign material. In this regard, it is fundamentally similar to the procedure described in 1995 by Fraser and Mee³ and in the same year by Brawn and associates⁴ of Birmingham, United Kingdom. Both groups have achieved noteworthy results with respect to early survival, and the 83% hospital survival in the present report compares very favorably with most contemporary results. However, a careful analysis of the results presented by Poirier and coworkers² suggests that this operative method may be less suitable for patients with the tiny ascending aorta that is the hallmark of aortic atresia, the most extreme and most common anatomic subset of HLHS.

The number of patients in this report with a truly diminutive ascending aorta (less than 2.5 mm) corresponds very closely to the number with aortic atresia. They make up a bit more than one half of the total number of patients with HLHS, and the rates of early and intermediate mortality for this group are 33% and 50%, respectively. In recent cases, the authors appropriately addressed the challenge of ensuring adequate coronary blood flow through a series of three modifications to the operative technique and expressed optimism with respect to the procedure that has most recently evolved. Yet early and intermediate mortality are not significantly lower in the latter half of the series than in the first half. By way of contrast, aortic atresia and small size of the ascending aorta have not been identified as risk factors for mortality in series from Philadelphia⁵ and Ann Arbor,⁶ wherein the technical features of stage I palliation included lengthwise incision into the ascending aorta and patch augmentation of the aortic reconstruction. Other series with other operative techniques have identified aortic atresia and small aortic size as risk factors for mortality and have been correlated with autopsy findings of myocardial ischemia and pre-coronary stenosis.^{7,8}

The palliated state of patients with HLHS is a period of significant vulnerability. This is certainly in large part a consequence of the abnormal physiology of coronary blood flow in this state. There is considerable diastolic runoff into the systemic–pulmonary artery shunt. Fogel and associates⁹ used echocardiography and Doppler flow studies to assess coronary blood flow in patients with aortic atresia. After stage I palliation, coronary blood flow was predominantly during systole, as compared with the predominance of diastolic coronary blood flow in normal hearts. In addition,

coronary blood flow requirements in the palliated state considerably exceed normal levels until the second-stage procedure, when the volume load on the single ventricle is reduced. Thus, the merit of technical modifications to the Norwood stage I palliative procedure must be considered with respect to their influence on coronary blood flow and its impact on survival, not only at the time of initial palliation, but throughout the several months of life that precede the second-stage procedure.

Intuitively, it is attractive to assume that the elimination of patch material in an aortic reconstruction may be associated with a lower incidence of late aortic arch obstruction. Nonviable patch material, even homograft tissue, is not expected to grow. Curiously, nearly all aortic arch obstruction after initial palliation of HLHS is encountered in the first several months of life.¹⁰ This is almost certainly related to the technical adequacy of the reconstruction rather than non-growth of the augmented aorta. Growth characteristics of the reconstructed aortic arch after surgical treatment of HLHS have been studied by Mahle and colleagues¹¹ in patients operated on a decade ago by Dr Norwood. These echocardiographic studies demonstrate that growth of the reconstructed aorta parallels growth seen in normal subjects. Postmortem investigations in Mahle's study reveal that all of the growth of the reconstructed aorta occurs in the native tissue that makes up at least a portion of the circumference of the aorta at every level. In angiographic studies done on patients 4 to 10 years after initial palliation, the aortic arch looks like an aortic arch, although larger than normal proximally. At present, one can only speculate that similarly satisfactory results will eventually be observed in patients whose aortic reconstruction uses only autologous tissue.

Although it is essential that we continue to refine the strategies of operative management of HLHS, at the heart of this issue is the question of whether Norwood's stage I palliative operation should be altered in ways that may introduce new variables with respect to coronary perfusion, particularly in patients with aortic atresia and a truly diminutive ascending aorta. It is conceivable that the most recent modifications of aortic configuration described by Poirier and associates² may eliminate the problems with coronary insufficiency that were encountered early in the series. It is important that technical advances continue to be made both with respect to preoperative, intraoperative, and postopera-

tive management of the abnormal physiologic state of these patients and with respect to the surgical exercises themselves. In the latter area, modifications such as those proposed by Poirier and coworkers² have among their objectives the goal of reducing the potential effects of cardiopulmonary bypass and circulatory arrest. Further progress in these areas is expected to contribute to the optimization not only of survival, but also of functional and neurologic outcomes. Growth of the surgically created pathways remains a consideration and will be the subject of ongoing investigations. The lessons learned in the surgical management of HLHS not only have dramatically affected the lives of children with this common heart malformation, but also have contributed immensely to the successful management of the entire spectrum of critical heart malformations necessitating surgical intervention early in life.

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