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Indications, criterions, and principles for biventricular repair

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THE HYPOPLASTIC LEFT HEART SYNDROME constitutes a wide spectrum of cardiac malformations, characterized by varying degrees of underdevelopment of the structures of the left heart and aortic arch. At the severe end of the spectrum, there is aortic and mitral atresia, and the left ventricle is vestigial. In contrast, at the mild end, there is hypoplasia of the left ventricle in the absence of overt stenosis of either the aortic or mitral valves, the combination we have described as hypoplastic left heart complex.[1] Until recently, debate with regard to the optimal surgical approach has centered on the choice between the Norwood operation and neonatal cardiac transplantation. In the last several years, nonetheless, it has been shown that of biventricular repair can be achieved in those patients at the favorable end of the spectrum that we designated as having the complex rather than the syndrome.[1,2] It is well recognized that an ongoing difficulty is the inconsistent and imprecise definition of hypoplastic left heart syndrome. In this presentation, I summarize our own criterions, and our principles for biventricular repair, as based on our experience at the Montreal Children's Hospital, for those patients we judge to have the hypoplastic left heart complex.

Anatomic substrates of hypoplastic left heart syndrome

The traditional anatomic substrates of hypoplastic left heart syndrome have been the combinations of aortic and mitral atresia; aortic atresia and mitral stenosis; aortic stenosis and mitral atresia; and combined aortic and mitral stenosis. To this list can be added critical aortic stenosis with hypoplasia of the left ventricle, congenital mitral stenosis with comparable left ventricular hypoplasia, the so-called hypoplastic left heart complex,[3] and severely unbalanced atrioventricular septal defect with hypoplasia of the left ventricle. Although the majority of patients with hypoplastic left heart syndrome have an intact ventricular septum, some may have a small-to-moderate ventricular septal defect. On the other hand, even though patients with aortic atresia, a large ventricular septal defect, and well developed left ventricles, have often been included in databases along with hypoplastic left heart syndrome for lack of a better place, they should not be considered in this fashion. Similarly, patients with other anomalies, such as double-outlet right ventricle with left ventricular hypoplasia, tricuspid atresia with discordant ventriculo-arterial connections, and double inlet left ventricle with or without obstructions of the aortic arch obstruction, should not be included within the hypoplastic left heart syndrome.

Pathophysiology

At the severe end of the spectrum, such as in patients with aortic and mitral atresia, the entire systemic circulation is dependant on flow from the right ventricle through the right ventricle to the patent arterial duct, with retrograde flow in the aortic arch and ascending aorta, the latter essentially serving as a coronary arterial conduit. At the mild end of the spectrum, in contrast, as in those with the hypoplastic left heart complex, or in critical aortic stenosis with left ventricular hypoplasia, the systemic circulation may be only partially dependant on flow through the arterial duct. In this setting, there is forward flow through the left ventricle into the ascending aorta and the aortic arch and its branches, with only the flow to the circulation fed through the descending aorta being duct dependent. Indeed, in some of these patients, the systemic circulation may be entirely dependent on the left ventricle, even in the presence of a patent arterial duct. In these patients, nonetheless, the closure of the patent duct may precipitate severe congestive heart failure

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due to the multiple obstructions found in the left heart and aortic arch.

Biventricular repair for hypoplastic left heart syndrome

Three surgical options have now evolved for patients with hypoplastic left heart syndrome:

- a multistage surgical reconstruction based on functionally univentricular physiology, as popularized by Norwood et al.[4];
- cardiac transplantation as advocated by Bailey and colleagues[5]; and
- biventricular repair as reported by our group.[1,2]

While the first two approaches can be applied to any patient with hypoplastic left heart syndrome, the biventricular approach can only be used in limited patients who fulfill careful criterions of selection. Since our purpose in producing this review is to discuss the biventricular approach, we will not discuss further our own experience with the Norwood operation, nor neonatal cardiac transplantation.

Until recently, biventricular surgical repair was entertained only for patients with critical aortic stenosis. In this respect, Rhodes and associates[6] established retrospective criterions to establish the left ventricular size that would favor biventricular repair in patients with critical aortic stenosis. They concluded that the adverse effects of a small inflow, a small outflow, and a small left ventricular cavity size, are cumulative. It is clear from their study, nonetheless, that using the previously suggested value of 20 ml/m^2 for left ventricular volume is not very predictive of outcome. Van Son and associates[7] have already questioned the predictability of biventricular repair using these previously defined criterions for ventricular volume in patients with right ventricular volume overload, such as those with right dominant unbalanced atrioventricular septal defects. Recently, in the largest multi-institutional study on critical aortic stenosis, conducted by the Congenital Heart Surgeons' Society,[8] a regression equation was calculated to help make the decision between univentricular and biventricular repair.

At the same time, our group has reported our efforts to achieve biventricular repair in a group of patients having other than critical aortic stenosis at the favorable end of the spectrum of hypoplastic left heart syndrome.[1,2] These patients were all judged to have hypoplasia of the components of the left heart and aortic arch, but in the absence of overt stenosis of the aortic and mitral valves. We dubbed this combination the hypoplastic left heart complex.[3] At the same time, several other authors[9–14] have reported biventricular repair patients with hypoplasia of the left ventricle without aortic stenosis, and with subsequent growth of the left ventricle after repair. Had we seen these patients, we would have labeled them as having hypoplastic left heart complex.

The surgical approach for such patients differs from those with critical aortic stenosis, in that the aortic valve, although hypoplastic, is not intrinsically stenotic, and does not require surgical valvotomy. This also suggests that biventricular repair may be possible in patients with a greater degrees of hypoplasia of the aortic and mitral valves and the left ventricle. The fact that the criterions suggested by Rhodes and colleagues[6] for patients with critical aortic stenosis do not apply to patients with hypoplastic left heart complex has recently been endorsed by Tani and associates.[13] Furthermore, Foker and associates[15] have induced rapid ventricular growth in infants we would consider to have hypoplastic left heart complex, or those with severely unbalanced complete atrioventricular septal defect, by first reducing the dimensions of the atrial septal defect with a snare, and then banding the pulmonary trunk. Biventricular repair was then achieved once there had been adequate growth of the left ventricle.

Biventricular repair for hypoplastic left heart complex

The decision of whether to consider biventricular repair in patients with hypoplastic left heart complex is based not only on the echocardiographic assessment of the size and function of the components of the left heart and aorta arch, but more importantly on the combined physiological consequence of the hypoplasia of these structures. The criterions and principles for biventricular repair, as developed and used at The Montreal Children's Hospital[1,2] are as follows:

- there must be antegrade flow of blood through the left heart into the ascending aorta and the proximal branches of the aortic arch;
- there must be no intrinsic stenosis of the aortic and mitral valves, the obstruction in the left heart existing by virtue of hypoplasia of the valves in keeping with the left ventricular hypoplasia;
- there should be adequate left ventricular function and no endocardial fibroelastosis.

If these physiologic and anatomic criterions are fulfilled, it is our belief that the particular size of each structure in the left side of the heart is less important. The size of the extracardiac structures, such as the ascending aorta and the aortic arch, are also irrelevant, since they can be easily enlarged to the desired size during the surgical reconstruction.

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Surgical principles and technique of biventricular repair

The principles of biventricular repair for patients with hypoplastic left heart complex are as follows:

- the elimination of the extracardiac anatomical afterload by enlargement of the aortic arch and ascending aorta down to the aortic root by insertion of a patch fashioned from a pulmonary homograft;
- the full preloading of the left heart, and elimination of the intracardiac shunts, by closure of the atrial septal defect, as well as the ventricular septal defect if present;
- an initially conservative approach to the aortic and mitral valves and the left ventricular outflow tract.

Patients undergo repair through a median sternotomy, with the use of cardiopulmonary bypass for core cooling to deep hypothermia. Although early in our experience we used deep hypothermic circulatory arrest, more recently we have used low-flow antegrade cerebral perfusion,[16–18] a technique described more fully elsewhere in this Supplement (see pp 70–74). With the patient under deep hypothermia, the aortic cannula is advanced into the brachiocephalic artery and snared in place. The proximal ends of the brachiocephalic, left common carotid, and left subclavian arteries are snared, and the upper descending thoracic aorta is clamped. Under antegrade cerebral perfusion at low rates of flow, the undersurface of the aortic arch is opened longitudinally from the proximal ascending aorta, past the insertion of the duct, and into the upper part of the descending thoracic aorta. The opened aorta is then enlarged with an appropriately fashioned patch made from a pulmonary arterial homograft. Any interatrial communication is closed through a right atriotomy. At this point in time, the need for atrial fenestration is unclear. Any ventricular septal defect, if present, is also closed.

The experience at Montreal Children's Hospital

Since November 1988, we have repaired 14 patients with hypoplastic left heart complex. Biventricular repair was achieved in 13 patients (92.8%), with the Norwood operation being performed in one. In 5 patients, there was a ventricular septal defect, while 1 patient had a right ventricular dominant atrioventricular septal defect with shunting exclusively confined at atrial level. The oval fossa was deficient in 7 patients, and probe-patent in 5.

Following biventricular repair, infusion of epinephrine was usually needed to permit discontinuation of cardiopulmonary bypass. During the immediate postoperative period, the left atrial pressure was high, but had decreased to almost normal levels within one or two postoperative days. It proved necessary to delay closure of the sternum in the majority of patients. The components of the left heart had enlarged significantly at the time of hospital discharge, as previously published.[1] Of the 13 patients undergoing biventricular repair, the left heart was able to support the systemic circulation in 12 (92.3%), albeit that two patients died (14.3% mortality). One patient died intra-operatively because the left heart could not support the systemic circulation. The second patient died 14 days post-operatively from junctional ectopic tachycardia. Another patient died 39 months post-operatively following an attempted modified Konno operation. This patient also had pulmonary hypertension and a single right coronary artery.

The 11 early survivors undergoing biventricular repair have needed a total of 10 unplanned reoperations. Those procedures were required because of obstruction in the left ventricular outflow tract obstruction in 5 patients, two of whom underwent a second re-operation. In two other patients, a total of 3 re-operations were needed because of recurrent coarctation.

All 11 patients surviving are currently well from a cardiac point of view. Their functional status is in Classes 1 or 2 of the grading system of the New York Heart Association. In the patient submitted to a primary Norwood operation, we have now performed successfully a bidirectional cavopulmonary anastomosis followed by an extracardiac Fontan operation.

Conclusions

Biventricular repair can be performed successfully in most patients falling in the malformation we dubbed the hypoplastic left heart complex.[1,2] Although growth of the components of the left heart has been observed by the time of hospital discharge, reoperation because of obstruction in the left ventricular outflow tract has been needed in almost half the patients. Only additional long-term follow-up to establish the outcome and functional status of the patients will ultimately determine whether the biventricular approach is optimal for those at the milder end of the hypoplastic left heart syndrome.

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