Hybrid approach to the comprehensive stage II operation in a subset of single-ventricle variants

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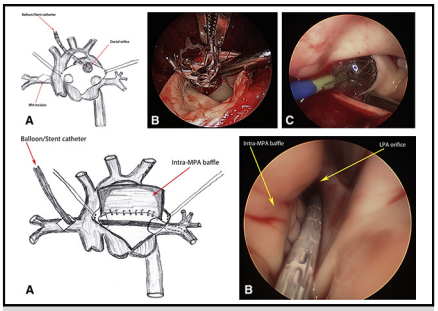
ABSTRACT

Objective: The objective of a hybrid approach to staged palliation of single-ventricle anomalies is designed to minimize the trauma of the first stage. However, the second stage is a complex procedure that may negate the advantages of the first stage. We sought to devise a ''hybrid'' approach to the second stage when aortic outflow is expected to remain unobstructed.

Methods: The procedure involves a simple incision into the main pulmonary artery, dilation/stenting of the ductal continuation, formation of a stented baffle between the branch pulmonary arteries' orifices, and a bidirectional Glenn connection. It avoids dissection of the distal arch and ductal continuation and obviates the need for a Damus-Kaye-Stansel connection. We carried out this procedure in 2 patients, one with unbalanced atrioventricular canal and the other with mitral atresia.

Results: Both patients underwent an uncomplicated operative procedure. Both patients were successfully weaned from the ventilator, with no clinically evident neurologic injury. The first patient died of complications related to thrombosis of the left pulmonary artery before initiation of anticoagulation. The second patient is alive and well 1 year postoperation with no obstruction to either systemic or pulmonary flow and no baffle leak and good right ventricle function.

Conclusions: This hybrid comprehensive stage II operation appears feasible and technically simpler than the conventional comprehensive stage II procedure. It is applicable to a subset of single-ventricle cases in which aortic outflow is anticipated to remain unobstructed. We recommend early postoperative anticoagulation to avoid early left pulmonary artery thrombosis. (J Thorac Cardiovasc Surg 2015;149:1095-100)



Intramain pulmonary artery baffle in hybrid comprehensive stage II procedure.

Central Message

This hybrid comprehensive stage II operation is applicable to a subset of single-ventricle cases in which aortic outflow is anticipated to remain unobstructed. It appears feasible and technically simpler than the conventional comprehensive stage II procedure. We recommend early postoperative anticoagulation to avoid early left pulmonary artery thrombosis.

Perspective

The objective of a hybrid approach to staged palliation of single-ventricle anomalies is to minimize the trauma of the first stage. However, the second stage is a complex procedure with potential complications that may negate the advantages of the first stage. We present a hybrid approach to the second stage in a subset of patients with sufficient antegrade aortic outflow to sustain upper body perfusion. The technique is less complicated than the conventional comprehensive stage II operation, avoiding distal arch dissection and reconstruction and an aortopulmonary anastomosis and with further technical developments may be achieved without hypothermia or cardiac and circulatory arrest.

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The ''hybrid Norwood'' sequence of staged palliation is a potential alternative to the conventional surgical management of patients with hypoplastic left heart syndrome (HLHS) and single-ventricle variants. The procedure consists of bilateral branch pulmonary artery banding, ductal stenting, and, when indicated, balloon atrial septostomy. The procedure is typically performed without cardiopulmonary bypass. The putative advantage of this approach is the avoidance of major surgical trauma in the newborn period. Although some centers report laudable outcomes for the hybrid approach, others report intermediate-term outcome (mortality) no better than that of the conventional staged palliation.[1-5] One study found no difference in neurodevelopmental outcome at 1 year between patients managed with the conventional approach and those managed with the hybrid approach.[6]

Although the first stage of the hybrid approach avoids a prolonged operation with cardiopulmonary bypass, deep hypothermia, and circulatory arrest, these techniques are typically required at the time of the ''comprehensive stage II'' operation. Additionally, this operation requires management of the ductal stent and, in some cases, bilateral

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Abbreviations and Acronyms

AV = atrioventricular
DILV = double-inlet left ventricle
DKS = Damus-Kaye-Stansel
HLHS = hypoplastic left heart syndrome
LPA = left pulmonary artery
LV = left ventricle
MPA = main pulmonary artery
PA = pulmonary artery
PDA = patent ductus arteriosus
PG = Palmaz Genesis
R = right
RPA = right pulmonary artery
RV = right ventricle
S = superior
TA = tricuspid atresia
VA = ventricular-arterial

pulmonary artery reconstruction because of band-induced trauma. The learning curve is known to be steep.[5] It remains a leap of faith that this deferral of trauma will result in better long-term survival and neurodevelopment for the patient.

To further reduce the total trauma to the patient, we propose an alternative technique to the standard comprehensive stage II operation, applicable to the subset of patients with sufficient antegrade aortic flow to sustain upper body perfusion. The technique avoids the creation of a Damus-Kaye-Stansel (DKS) connection and surgical arch reconstruction entirely and, with further technical development, may be achieved without hypothermia, cardiac and circulatory arrest.

PATIENTS

Case 1

The patient was a full-term 3.3-kg newborn with Down syndrome. Echocardiography demonstrated an unbalanced complete atrioventricular (AV) canal defect. The left ventricular (LV) volume index was 14 mL/m^2. The LV/right ventricle (RV) effective valve area ratio was 0.42 (AV valve index 0.29). The LV outflow track was not obstructed, but the aortic arch was hypoplastic, with a midtransverse arch–to–ascending aortic diameter ratio of 0.34. The isthmus was also hypoplastic and there was a large ductal arch. On day 4, the neonate underwent the hybrid stage I procedure. Following hospital discharge, echocardiography demonstrated that the ductal stent had migrated somewhat into the descending aorta, but the ductus remained patent with a gradient less than 10 mm Hg. At 5 months (7 kg), catheterization demonstrated unobstructed antegrade aortic flow to the arch vessels but no appreciable flow from the ascending aorta into the descending aorta. The aortic annulus, aortic root, ascending aorta, and transverse arch diameters were 8 mm (z = −0.9), 14 mm (z = +1.7), 12 mm (z = +1.2), and 6 mm (z = −1.9), respectively. The mean systolic gradient through the ductus into the descending aorta was 12 mm Hg. Mean distal left and right pulmonary artery (LPA and RPA, respectively) pressures were 24 and 10 mm Hg, respectively. The patient underwent the following hybrid stage II procedure:

Technique

Through a median sternotomy, we mobilized the heart, great vessels, and innominate and superior caval veins and divided the azygous vein. We did not mobilize the distal arch, aortic isthmus, or descending aorta. We placed dual arterial cannulas in the innominate artery and pulmonary artery (PA) trunk and venous cannulas in the right atrium and (later) innominate vein, then commenced cardiopulmonary bypass. After cooling to 18°C degrees, we clamped the ascending aorta, arrested the heart, and then removed the polytetrafluoroethylene bands from the branch pulmonary arteries. We then occluded the arch branches, removed the PA cannula, and commenced antegrade regional cerebral perfusion. (Occlusion of the arch branches may not be necessary if the distal arch is already discontinuous or if one can dilate or re-stent the ductal arch without a blood-free descending aorta. In this case, aortic, rather than innominate artery, cannulation can be performed, with perfusion to the entire upper body.) We did not occlude

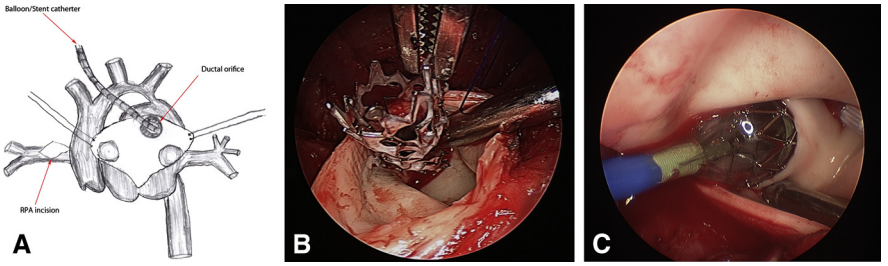

FIGURE 1. After commencing cardiopulmonary bypass with 1 or 2 arterial cannulas and a right atrium cannula, the branch pulmonary arteries are debanded. A, Under selective cerebral perfusion, an incision is made into the RPA at the proposed site of superior vena cava connection, usually at or just distal to the site of prior banding. Then, a longitudinal incision is made into the large main pulmonary artery. The location of the branch pulmonary artery and ductal orifices are noted. B, A protruding end of the original ductal stent is trimmed back. C, The ductal stent is then dilated under direct vision, and a second stent placed as needed and position determined by videoscope. *RPA*, Right pulmonary artery.