

# Bidirectional Cavopulmonary Shunt for Cyanotic Heart Disease: Surgical Experience from a Developing Country

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## Abstract

**Objective:** The importance of bidirectional cavopulmonary anastomosis for palliation of complex cyanotic congenital heart disease is widely recognized. This study was conducted to highlight our surgical experience with this procedure in a developing country.

**Methods:** A retrospective study was conducted using medical records at the Aga Khan University Hospital, Karachi, Pakistan. Clinical findings at presentation, anatomical defects seen on transthoracic echocardiography, pre-operative McGoon index, cardiopulmonary bypass time, use of cardioplegia, post-procedure oxygen saturations and complications were evaluated.

**Results:** A total of 8 patients underwent bidirectional cavopulmonary anastomosis. There were 6 males (75%) and 2 females (25%). Ages and weights at operation averaged  $5.7 \pm 3.7$  years (range 2-14 years) and  $18.6 \pm 10.4$  kg (range 8.5-35.5 kg) respectively. The most common symptoms were the presence of cyanosis in all (100%) patients followed by recurrent respiratory tract infections in 3 (37.5%) patients. Transthoracic echocardiography revealed 6 (75%) patients with atrial septal defects, 5 (62.5%) with tricuspid atresia, 3 (37.5%) with ventricular septal defects, 3 (37.5%) with malposition of great vessels, 2 (25%) with pulmonary stenosis and 2 (25%) with double inlet left ventricles. The mean pre-procedure McGoon index was  $2.1 \pm 0.5$  (range 1.37-2.80). All patients received cardioplegia. Cardiopulmonary bypass was used in all patients for a mean time of  $154.1 \pm 83.6$  minutes (range 60-298 minutes). All patients were ventilated for a mean period of  $1.5 \pm 0.7$  days (range 1-3 days). The ICU stay was  $3.0 \pm 0.6$  days (range 2-5 days) with a total hospital stay of  $9.8 \pm 3.8$  days (range 7-18 days). The mean post-procedure oxygen saturation was  $82.6 \pm 3.5\%$  (range 76-86%). The most common post-operative complication was supraventricular arrhythmia in 2 (25.0%) patients. There were no intra-operative or early (within 7 days of procedure) deaths. One patient developed pulmonary artery hypertension and died 23 months later due to cardiac arrest.

**Conclusion:** Patients tolerated the procedure well. After a mean follow-up of 10 months, 6 patients were assessed to be in New York Heart Association (NYHA) functional class I and one patient in NYHA class II. Clinical and post-procedural data gathered from our experience confirms the safety of bidirectional cavopulmonary anastomosis (JPMA 53:506;2003).

## Introduction

Fontan and Baudet reported the first clinically successful procedure for total bypass of the right side of the heart for treatment of tricuspid atresia.<sup>1</sup> Since the original description, a number of modification to Fontan's operation have been described that have simplified the operation and allowed its application to a variety of complex congenital heart lesions.

The bidirectional cavopulmonary shunt may be defined as an operation that diverts the systemic venous return from the superior vena cava or cavae to both lungs. The role of bidirectional cavopulmonary anastomosis for palliation of complex cyanotic congenital heart disease is widely recognized.<sup>2-4</sup> The results of Fontan operation appeared to be somewhat better when applied to patients with tricuspid atresia than to those with other defects.<sup>5,6</sup> The trend toward, early application of orthoterminal procedures<sup>7</sup> supports the concept of early indication for bidirectional cavopulmonary anastomosis as a safe and effective preparatory stage for a Fontan procedure. Here, we

report our 3-year experience with bidirectional cavopulmonary anastomosis.

## Materials and Methods

The study group included all patients who underwent bidirectional cavopulmonary anastomosis for palliation of cyanotic heart disease between January 1999 to December 2001. Hospital records were evaluated for clinical findings at presentation, previous palliative procedure and pre-operative transthoracic echocardiography (TE). TE was chosen as tool to assess ventricular function, anatomical defects (for diagnosis), valve status, and pericardial effusion.

Operative notes were evaluated for pre-operative McGoon index, cardiopulmonary bypass time, use of cardioplegia, any associated procedure beside bidirectional cavopulmonary anastomosis, and post-procedure oxygen saturation.

The standard surgical technique is described here. Through a standard median sternotomy, cardiopulmonary bypass with moderate to deep hypothermia was performed. Cardiopulmonary bypass was instituted by distal aortic cannulation and placement of two separate caval cannulas or, alternatively, a single atrial cannula, depending on the perfusion strategy. Previous systemic-pulmonary shunts were taken down (except in two cases). The superior vena cava (SVC) was transected and anastomosed end to side to ipsilateral pulmonary artery (Figure). The azygos vein was ligated primarily in all patients.

The variable in post-operative periods included: length of hospital stay (intensive care, special care, general care), length of assisted ventilation, and any complication.

## Results

Eight patients underwent bidirectional cavopulmonary anastomosis for palliation of cyanotic heart disease between January 1999 to December 2001. There were 6 males (75%) and 2 females (25%). Age and weights at operation averaged  $5.7 \pm 3.7$  years (range 2-14 years) and  $18.6 \pm 10.4$  kg (range 8.5-35.5 kg) respectively. The presenting complaints are enumerated in table 1. The characteristics of patients, diagnosis, procedures and follow-ups are given in table 2.

The mean pre-procedure McGoon index was  $2.1 \pm 0.5$  (range 1.37-2.80). Cardiopulmonary bypass was used in all patients with mean time of  $154.1 \pm 83.6$  minutes (range 60-298 minutes). All patients were ventilated with mean of  $1.5 \pm 0.7$  days (range 1-3 days). The ICU stay was  $3.0 \pm 0.6$  days (range 2-5 days) with total hospital stay of  $9.8 \pm 3.8$  days (range 7-18 days).

There were no intraoperative or early (within 7 days of procedure) deaths. One patient developed pulmonary artery hypertension and died 23 months later due to cardiac arrest. The post-procedure complications are shown in table 3.

Follow-up was available in all 8 patients. At a mean follow-up of 10 months, 6 patients are in New York Heart Association (NYHA) functional class I and one patient is in class II.

## Discussion

Over the past years, bidirectional cavopulmonary anastomosis has been increasingly used in the treatment of less than ideal candidates for a Fontan operation, usually as an intermediate stage after a systemic-pulmonary shunt performed in the first months of life.<sup>2-4</sup> Avoidance of bidirectional cavopulmonary anastomosis as primary palliation at an early age is mostly justified by previously reported failures with the classic Glenn anastomosis in younger children.<sup>8</sup> However, the current trend toward early orthoterminal repair and the concern aroused by the deleterious effects of systemic-pulmonary shunts on pulmonary arterial anatomy and on single ventricular function prompted us to consider bidirectional cavopulmonary anastomosis even for patients under 5 years of age.

Follow-up studies after the Fontan approach have shown that important risk factors for death or take down include ventricular function, especially compliance, pulmonary vascular resistance, and pulmonary arterial anatomy -often distorted by prior shunt procedures.<sup>9</sup> In contrast to a systemic-pulmonary shunt, the bidirectional cavopulmonary shunt does not increase ventricular work (thereby avoiding further ventricular hypertrophy and decreased compliance), does not put the pulmonary vascular resistance at risk, and does not distort the pulmonary arteries. In our series, all patients had good ventricular Demonstration of the theoretical hemodynamic advantages of bidirectional cavopulmonary shunt is extremely difficult because there is great variability in preoperative ventricular function and pulmonary vascular resistance among patients. Whereas previous reports on the Glenn shunt<sup>10</sup> or the Fontan procedure<sup>9</sup> discuss the importance of normal pulmonary vascular resistance, we believe that low pulmonary artery pressure, in addition to normal pulmonary resistance, is a critical success factor because elevated pulmonary artery pressure would cause cavopulmonary shunt to function improperly. The mean pulmonary artery pressure in our series was satisfactory. Because the pulmonary artery pressure is critical in determining outcome after cavopulmonary shunt, we recommend aggressive attempts at direct (through systemic-pulmonary shunts) or indirect measurement (using the pulmonary venous wedge pressure<sup>11</sup>). In our series, 3 patients had Blalock-Taussig shunt prior to cavopulmonary shunting. It has been shown earlier that, a 4 mm modified Blalock-Taussig shunt performed in the early stages of life will provide adequate palliation for about one year.<sup>12</sup>

We never considered a valvuloplasty at the time of bidirectional cavopulmonary anastomosis, speculating on a theoretical partial regression of valvular incompetence after normalization of ventricular volume load.

One of the important risk factors for death or takedown of the Fontan operation is dimensions of the right and left pulmonary arteries, expressed as McGoon index.<sup>13</sup> The risk of combined event increased sharply when McGoon index was less than 1.8.<sup>13</sup> In our series, mean McGoon index was 2.1.

There are several technical advantages to cavopulmonary shunt when compared with other Fontan modifications. The construction of a lateral tunnel avoids suture placement near the atrioventricular conduction pathway, decreasing the likelihood of postoperative heart block. The posterior location of the pulmonary artery anastomosis avoids the risk of sternal compression associated with other types of atriopulmonary anastomosis.<sup>14,15</sup> Pulmonary venous obstruction by the intraatrial baffle, which can occur with mitral atresia, is also avoided by the lateral tunnel.

The post-operative course after bidirectional cavopulmonary anastomosis has by comparison been benign, with no early deaths. The patients were ventilated for mean duration of 1.5 days which was comparable with earlier studies reporting median of 1 day.<sup>1,16</sup> The need for slightly prolonged ventilation in some of the patients was excessive bleeding. The ICU and hospital stay was also found to be consistent with previous studies.<sup>1,16</sup>

Atrial arrhythmias are common late after Fontan operation, reported to occur in more than 35% of patients.<sup>17,18</sup> Three patients (25.0% ) had supraventricular tachyarrhythmias and one patient (12.5%) had bradycardia in our series. These arrhythmias may result from the atriotomy and its closure, the suture line of the intraatrial baffle, as well as from the elevated pressures to which the right atrium is exposed.

Although we did not run any statistics owing to small number patients; however, clinical and functional information from our experience confirms the safety of bidirectional cavopulmonary anastomosis and its efficacy in providing adequate oxygenation. The operation can be performed with low morbidity and mortality and may result in improved hemodynamics when compared with other Fontan-type procedures. Further experience with this approach might culminate, in selected cases, with single-stage fontan repair.

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