

Survey of Fontan and Total Cavopulmonary Connection in Patients with Single Ventricle

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Abstract

Background- In recent decades, advances in congenital heart surgery have dramatically increased the survival of infants with critical congenital heart disease.

Methods- To determine the outcomes after the Fontan and total cavopulmonary connection (TCPC) procedures, we investigated 51 patients with single ventricle who underwent the Fontan (27 cases) and TCPC (24 cases) procedures between 1991 and 2001.

Results- Five patients (9.8%) died after surgery. Forty-six patients were followed for a period of 42.9 ± 32.1 months. Thirty-eight of them (82.6%) were alive with a better New York Heart Association functional classification than preoperatively. Ejection fraction was more than 60% in 28 cases (84.4%) one year after the operation. Complications were seen in 23 patients after surgery: 8 with early complications, 12 with late complications and 3 with both early and late complications. Overall arrhythmia was detected in 34.8% of our patients during their follow-up. Reoperations were necessary for 15.2% of our patients.

Conclusions- The surgical approach to congenital heart defects continues to involve a series of complicated operations with significant associated mortality and morbidity (*Iranian Heart Journal 2004; 5(3):23-29*).

Key words: Fontan procedure ■ total cavopulmonary connection ■ single ventricle ■ cardiac surgery

Within the past years, several experimental and clinical studies have been published in the field of surgery for congenital heart defects. General agreement has already been reached on the indications for treating most of the congenital cardiac malformations in order to improve the natural history of the disease. However, strong disagreement exists regarding the timing and methods of treatment, either for congenital heart defects, for which the approach should be standardized after years of use, and even more when a new technique or a new approach is introduced to replace the existing ones.¹

Over the past two decades, advances in congenital heart surgery, pediatric

cardiology and intensive care medicine have dramatically increased the survival of infants with critical congenital heart disease. The group of patients that has perhaps benefited the most from this progress has been the single-ventricle population. Staged palliation culminating in the Fontan procedure has resulted in a decreasing mortality rate and an increase in the number of single-ventricle survivors. Many studies have focused on outcomes after the Fontan procedure in recent years. These reports demonstrate progressive improvement in early postoperative survival and intermediate and late postoperative outcomes due to surgical innovations, such as the lateral tunnel and extracardiac Fontan modifications and

fenestration, as well as technological improvements, such as modified ultrafiltration. Despite these improvements, significant morbidity remains after the Fontan completion.² The Fontan operation and its modifications have remained an important milestone in the surgical management of selected patients with complex cyanotic congenital heart disease. In the original description of his operation in patients with tricuspid atresia, Fontan observed that the reduction in right atrial pressure, which followed extubation, was accompanied by a clinical improvement in all cases and concluded that spontaneous respiration should be established as early as possible in these patients.³

Structural defects generally managed with a staged palliation include variations of single left ventricle and variations of single right ventricle. Children with single-ventricle anatomy generally undergo palliative surgery in the neonatal period, which will provide for unobstructed systemic blood flow, restrictive pulmonary blood flow, unobstructed pulmonary venous return, unobstructed interatrial communication and a minimum of atrioventricular valve regurgitation. Subsequent procedures are performed as the transition of the circulatory system to the total cavopulmonary connections (TCPC) with passive pulmonary and pulsatile systemic blood flow.

A superior cavopulmonary anastomosis is usually performed at 4 to 6 months to volume unload the single ventricle.

It is at 18 to 24 months that most centers perform the modified Fontan procedure to complete the TCPC.^{2,4,5}

The purpose of this study was to review a single-center experience that had evolved over a decade and to determine the outcomes after the Fontan and TCPC procedures.

Methods

This is a retrospective study over a 10-year period designed to ascertain the result of the Fontan procedure and total cavopulmonary connection in patients with single ventricle at one of the referral cardiothoracic surgery centers in Tehran, Iran.

Fifty-one patients (26 males and 25 females) within the 4 to 24 age bracket (median: 9 years) underwent the Fontan and TCPC between 1991 and 2001 in our center. This study was approved by our institutional ethics committee. Informed consent having been obtained from the patients before surgery, their records were reviewed and analyzed for findings before and after the operation. Data analysis was done using SPSS statistical software package (version 10.0).

Results

Characteristics of patients

Twenty-seven patients (11 males and 16 females) with a mean age of 8.3 ± 3.4 years (range: 4-15 years) underwent the Fontan operation, and 24 patients (15 males and 9 females) with a mean age of 11.3 ± 4.3 years (range: 4-24 years) had the TCPC procedure. The mean age at surgery did not differ in early experience (1991 to 1996) and late experience (1997 to 2001).

Congenital heart defects

The patients were operated on because of their congenital heart defects, namely tricuspid atresia, single ventricle, transposition of great arteries, double outlet right ventricle and hypoplastic left heart syndrome (Fig.1). In addition, 21 patients (41.2%) had associated cardiac anomalies, including atrioventricular valve regurgitation, right aortic arch, dextrocardia, left superior vena cava,

patent ductus arteriosus, single coronary artery, interrupted inferior vena cava, single coronary artery and right coronary artery with origin of left descending artery.

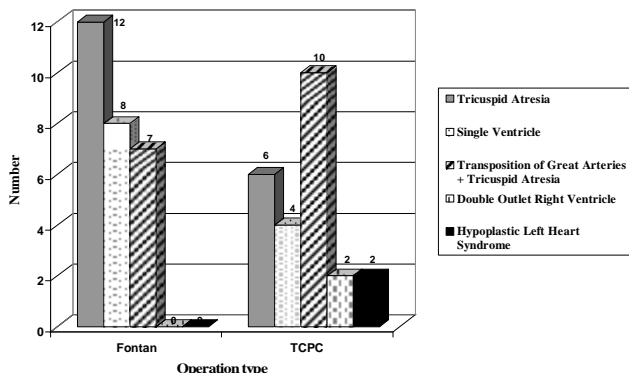


Fig. 1. Congenital heart defects of patients who underwent surgery.

Previous operation

Twenty-two patients had a history of prior cardiothoracic surgery before this admission, the most common being the Goretex shunt (15 cases). Also, the hemi-Fontan had been performed for two cases before this operation.

Operation

All the patients had normal sinus rhythm during the operation. The mean time of cross-clamp was 102.8 ± 57.4 minutes (40.7 ± 35.0 minutes for the Fontan and 65.6 ± 41.9 minutes for the TCPC).

Patients' status at discharge

O₂ saturation of 31 patients (67.4%) was more than 90% at the time of discharge from the intensive care unit. It was between 70%-90% in 13 patients (28.3%), and only 2 patients had O₂ saturation lower than 70%. The mean of central venous pressure was 14.9 ± 3.9 mmHg at the time of discharge from hospital (16.3 ± 3.3 mmHg after the Fontan and 13.6 ± 4.0 mmHg after the TCPC).

Patients' status at follow-up

Forty-six patients were followed for a period of 42.9 ± 32.1 months with 1887 patient-months of follow-up. The New York Heart Association (NYHA) functional class improved significantly after surgery (P value = 0.002). Forty-five out of 51 patients were in NYHA classes I and II, postoperatively; and 29 were in NYHA classes I and II, preoperatively. Thirty-eight of them (82.6%) were alive with a better NYHA functional classification than preoperatively. Considering class I alone, there was only one patient so classified preoperatively versus 27 so classified postoperatively (Table I).

Table I. NYHA functional class before and after surgery.

	Fontan				TCPC			
	Before operation		After operation		Before operation		After operation	
	Number	Percent	Number	Percent	Number	Percent	Number	Percent
Functional Class I	1	3.7	15	60	-	-	12	57.1
Functional Class II	15	55.6	9	36	13	54.2	9	42.9
Functional Class III	11	40.7	1	4	11	45.8	-	-

Ejection fraction was more than 60% in 28 cases (84.4%) one year after surgery (Fig. 2).

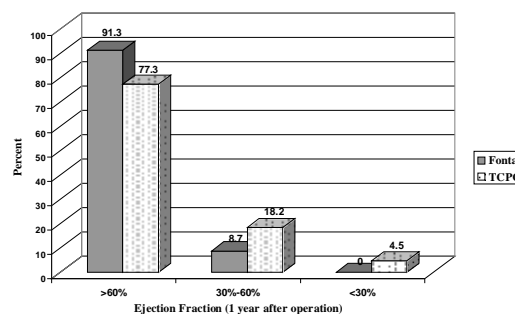


Fig. 2. Ejection fraction, one year after surgery.

Mortality after operation

Five patients (9.8%) died after the operation. Three of them died after the Fontan (11.1%) because of low cardiac output, systemic arterial and venous hemodynamic abnormalities and thromboembolic complications. The other two patients died after the TCPC (8.3%) because of low cardiac output and hemorrhage. These patients' age at surgery was 11.5 ± 3.8 years.

Complications after operation

Early and late complications after surgery were seen in 23 patients (12 after the Fontan and 11 after the TCPC): 8 with early complications, 12 with late complications and 3 with both early and late complications. The most common complications after surgery were pleural effusion (9 patients), pericardial effusion (5 patients), ascitis (4 patients), jaundice (4 patients), hemorrhage, chylothorax, pulmonary vein stenosis, brain abscess, inferior vena cava thrombosis, generalized edema, dyspnea and hemoptysis.

Arrhythmia occurred in 5 patients (10.9%) within 1 year after the operation (3 patients after the Fontan and 2 patients after the TCPC), including: right bundle branch block (3 cases), complete heart block (1 case) and junctional ectopic tachycardia (1 case). The incidence of arrhythmia increased with time, which was seen in 11 patients (23.9%) in the following years. Only 2 cases belonging to the group after the Fontan procedure were complicated with atrial flutter in the follow-up. Overall arrhythmia was detected in 34.8% of our patients during their follow-up (33.3% after the Fontan and 36.4% after the TCPC).

Reoperation

Seven patients (15.2%) underwent reoperation after this operation (6 days- 7 years): two during this admission (2 after

the TCPC) and five after discharge (2 after the Fontan and 3 after the TCPC).

Discussion

This study represents a single center experience evolving over a 10-year period on 51 patients, who underwent the Fontan and TCPC operations. Inter-institutional and intra-institutional differences in technique, timing, staging and variations in perioperative management make comparing studies on survival after the Fontan procedure problematic. Despite these confounders, analyses on the evolution of the Fontan over the past decade, from the original valve-less atriopulmonary connection to the staged total pulmonary cavopulmonary connection with fenestration, have shown a progressive and dramatic improvement in operative survival.² The surgical approach to congenital heart defects continues to involve a series of three or more complicated operations with significant associated mortality and morbidity.⁶

Age at surgery ranged from 4 to 24 years (mean 9.7 ± 4.1 years, median 9 years). Younger age at surgery was an independent risk factor for early failure. This finding has been reported previously by others,^{5,7} but the mechanism remains speculative. Possible contributing factors include smaller patient anatomy and a more reactive pulmonary vascular bed after cardiopulmonary bypass.⁵ Cetta F. et al., who had operated on 339 consecutive patients at the Mayo Clinic between 1987 and 1992, demonstrated that younger age at the time of surgery was one of the many factors which might have contributed to a decreased early mortality rate after the Fontan.⁸ Fishberger SB. et al. noticed that atrial flutter developed sooner and was more likely to occur in patients who were older at the time of the Fontan operation.⁹ Cecchin F. et al. found that older patients

at surgery were associated with a higher incidence of atrial and ventricular arrhythmias.¹⁰

The NYHA functional class was improved significantly after our operations. Functional Class I was reported in 58.9% of our patients after surgery, which was similar to a previous study conducted on 155 patients at Mayo Clinic by Mair DD. et al., who reported the prevalence of 56% for their patients in class I.¹¹ There are, of course, many ways to assess the functional status, the most common being the NYHA classification.¹² In a follow-up study by Fontan et al.,¹³ 48% of the patients were in NYHA class I, 16% were in class II, 2% were in class III and 33% were dead.¹³ Recent studies^{5,7,14} indicate that more than 90% of their patients were in NYHA classes I and II, postoperatively; our data (97.8%) were similar. All of our patients who underwent definitive TCPC were in NYHA classes I and II after surgery; the same figure was more than 76% in a study in Austria.¹⁵ When assessing the functional status with the NYHA classification, it is important to compare the patient's status after surgery with that before surgery. In our study, 82.6% of the patients were alive with a better NYHA functional classification than preoperatively; the figure stood at more than 34.7% in a previous study by Driscoll DJ. et al.¹² A poor functional outcome is uncommon after the Fontan operation.⁵

Mortality after the Fontan operation was 11.1%, which was similar to the figure in a previous study⁸ at the Mayo Clinic (9.1% of 339 patients) and more than 5.4% of 129 patients at James Whitcomb Riley Hospital for Children⁷ and 6.6% of 352 patients at the Children's Hospital of Philadelphia.¹⁶ The Fontan operation can be performed with a mortality risk of less than 10%. Late results are encouraging when contrasted with the clinical course of patients before this operative approach is utilized.¹¹

Although the Fontan operation can be carried out in many centers with an operative mortality of <5%, these figures apply only to those undergoing the Fontan, not to the entire cohort. Surgical mortality thus addresses the "tip of the iceberg".¹⁷ Mortality rate after our TCPC (8.3%) was lower than that in previous studies: 17% of 47 patients at a center in Austria,¹⁵ 15% of 40 patients in a study by Balaji S. et al.,¹⁸ and 10.5% of 76 patients in a study by Yoshimura N. et al.¹⁹ Mortality did not statistically differ between the Fontan procedure and TCPC, in agreement with a previous study.¹⁸

The causes of mortality in these groups were low cardiac output, systemic arterial and venous hemodynamic abnormalities, thromboembolic complications and hemorrhage. The operative mortality for the modified Fontan operation has been well defined for a large number of patients by numerous authors. As experience with this procedure has increased, the operative risk has decreased.¹² Despite a number of modifications in surgical techniques, favorably influencing the early postoperative course of patients undergoing Fontan-like operations,⁴ the chief cause of morbidity and mortality in this group remains a low cardiac output state, hemodynamic abnormalities and thromboembolic complications. A low cardiac output is not in itself a surprising feature in the immediate postoperative period, when the important influences of mechanical ventilation are superimposed on the inevitable global effects of cardiopulmonary bypass on the heart and lungs.²

Half of our patients experienced early and late complications after surgery, which were 23.9% and 32.6%, respectively. When analyzing the outcome studies performed today, one must remember that any outcome investigation reflects the management strategies used more than a

decade ago. The evolution in management that has occurred over the past two decades will almost certainly improve the outcomes in the future.² The report by Gaynor JW. et al.¹⁶ demonstrated that early survival after the modified Fontan has improved dramatically over the past decade. Similarly, the intermediate and late survival of Fontan patients has improved as well. The most common complications after our operations were pleuropericardial effusions, seen in 30.4% of the patients; this figure was lower than that in a study on 47 consecutive patients at a center in Austria in which 42% of their patients suffered from temporary pleuropericardial effusions.¹⁵

Arrhythmia was reported in 34.8% of our patients during their follow-up; it was higher than 20% in a study by Driscoll DJ. et al.¹² and 4.7% in a study by Bando K. et al.⁷ Two important studies from Boston and New York have shown that both staged approaches to the Fontan and TCPC are associated with sinoatrial node dysfunction early after these procedures.²⁰ Furthermore, atrial dysrhythmias and poor functional outcome have been major concerns during late follow-up.⁷ After the completion of the Fontan, arrhythmias result from sinus node dysfunction, increased atrial pressure and the presence of suture lines and scars. The incidence of atrial tachyarrhythmias and bradyarrhythmias increases with time. Much of the research on arrhythmias after the Fontan has focused on identifying the predictors for atrial arrhythmias or sinus node dysfunction, and many have focused on surgical strategy as a possible critical predictor.² In addition, it is important to recognize the presence of atrial flutter in patients who have had the Fontan operation because it may result in congestive heart failure and fluid retention.¹² Whereas the frequency of arrhythmias did not statistically differ

between the Fontan and TCPC in our study, Balaji S. et al. demonstrated that arrhythmias in the Fontan procedure were more frequent than those in the TCPC.¹⁸

Reoperations were necessary for 15.2% of our patients, which was much lower than the figure of 29.3% in a study by Driscoll DJ. et al.¹² The need for reoperation after the Fontan procedure was relatively frequent.¹²

Conclusion

Staged palliation culminating in the Fontan and TCPC procedures have resulted in a decreasing mortality rate and an increase in the number of single-ventricle survivors. There has been a progressive improvement in early postoperative survival and intermediate and late postoperative outcomes due to surgical innovations. Despite these improvements, significant morbidity is still seen after these operations. It is not clear how the advances of yesterday will affect the outcomes of tomorrow. The challenge is to strive through multi-institutional cooperative clinical research to attempt to answer the many questions that remain on outcomes in the single-ventricle population.²

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