

Fontan Operation: Reflections on its Current Evolution and Perspectives

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As the Fontan operation¹ became the most frequently performed surgery in Pediatric Cardiology for the functional correction of many congenital heart diseases with single-ventricle physiology, it started to be considered in relation to the most suitable maneuvers to minimize the complications that still challenge its management in the long term.

Although the incidence of such complications has decreased mainly as from 1988 with the introduction of the total cavopulmonary technique², we can still observe the occurrence of morbidity elements whose rates are disturbing and affect the evolution of this method.

Because of the dynamics imposed on the new anatomic situation with the cavopulmonary connection which eliminates the pulsatile function of the right ventricle and establishes the slow venous flow through the venae cavae to the pulmonary arterial tree, the occurrence of complications resulting from congestive, electrical and thromboembolic aspects is thus facilitated.

The congestive aspects are characterized by increased venous pressure, hepatomegaly, protein-losing enteropathy due to lymphangiectasia, lower-extremity edema, and ascitis.

The electrical aspects concern both the decrease in sinus automatism and supraventricular arrhythmias such as extrasystoles, fibrillation, and paroxysmal tachycardias.

The thromboembolic aspects, which start in the venous system and in venous-arterial junctions and conduit connections, are also related to the occurrence of several coagulation disorders caused by factors resulting from liver congestion.

Pathogenic factors of these adverse elements are related to preoperative anatomic and functional aspects and that is why a more accurate analysis of these aspects is mandatory at the moment of the surgical procedure.

In this setting, it is well timed to recall and mention the factors considered optimal for the surgical indication, which are mainly: preserved ventricular function close to normal, adequate pulmonary arterial tree with mean pressures below 15 mmHg and pulmonary resistance lower than 2U Wood, and also the presence of sinus rhythm responsive to stimuli³. Absence of atrioventricular and aortic regurgitation, of fistulae and systemic-pulmonary anastomotic vessels, in addition to myocardial hypertrophy are also included.

These deleterious factors are effectively responsible for the mortality that occurs throughout time and which ranges, according to the expertise of each medical center, from 15 to 30% 15 years after the surgical procedure³. That is where death results from, mainly related to heart failure, arrhythmias, thromboembolism, and to protein-losing syndrome.

Among the complications, those with the highest prevalence and repercussion are heart failure and arrhythmias, mainly in atriopulmonary⁴⁻⁶ techniques, but also in cavopulmonary⁷ procedures when performed in adulthood. These complications are less frequent in cavopulmonary connections with extracardiac conduits⁸ than in intra-atrial lateral tunnel^{7,9}. Other complications such as thromboembolism and protein-losing enteropathy are also directly related to the atriopulmonary technique^{4-6,10} and also when patients undergo surgery in adulthood⁶. These latter complications were less frequent in the cavopulmonary technique^{8,9,11,12}, mainly when the extracardiac conduit⁸ was used (Tab. 1).

Despite these findings, however, other assessments show that the outcome of patients undergoing surgery in adulthood is similar to that of patients undergoing surgery in childhood, as regards the time for the onset of complications and the actuarial curve of postoperative survival¹⁰.

Thus, according to Burkhart et al¹⁰, among 132 adult patients undergoing surgery between 18 to 53 years of age with an immediate mortality of 11 (8.3%) patients and survival of 68% after 15 years, the long-term complications, with a mean 9.1-year postoperative follow-up, were represented by protein-losing enteropathy in 8 patients (7.4%), pleural effusion in 7 (6.5%), stroke in 4 (3.7%), complete atrioventricular block in 16 (15%), and heart failure in 6 (5.2%), in addition to inferior vena cava thrombosis in 1 patient. By way of comparison, in childhood, according to Lee et al¹³, among 405 cases analyzed after a 10-year postoperative follow-up, thromboembolism occurred in 1.2%, arrhythmias in 17%, and heart failure in 2.4% of the cases.

Given the similar outcomes of these age groups, there is a growing uncertainty as to the optimal timing for surgery, as to whether it should be performed earlier or at its adequate natural timing.

Incidentally, in this setting, to further reinforce the controversy on the optimal timing for surgery, Lee et al¹³ have reported that the survival of the 405 patients undergoing surgery from 1988 to 2000, mean age of 28 months, was

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Point of View

	N	Mean Age (years)		Technique						Mortality	
		at operation	long term	HF	Arrhythmia	TE	PLE	AP	CP	Imm	Late
Cazzaniga M	124	7.3±4.7			33(34.7%)		5 (5.2%)	102	22*	29 (23%)	20(16%)
van den Bosch AE	36	12 (2-34)	15 (0-23)	14 (36.8%)	20(56%)	9(25%)		32	4*		10(28%)
Veldtman GR	61	24(18-47)	10	20 (34%)	34(57%)	14(32%)	-	51	9*	8 (13%)	10(17%)
Burkhart HM	132	18-53	9.1	6 (5.2%)	16 (15%)	5 (4.7%)	8 (7.4%)	110	19* 3**	11 (8.3%)	34(32%)
Mott AR	23	23(18-41)	30 m	3 (13%)	2				12* 11**	-	-
Kaulitz R	142	66.3± 57.9m	91.1± 43.9m			10 (7%)	4 (2.8)		121* 21**	-	-
Stamm C	220	11m-32 a	10.2± 0.6	26 (13%)	60 (30%)		3 (1.5%)		220*	12 (5.4%)	7(3.3%)
Tokunaga S	100	6.1±4.2	37.3m						100**	-	5(5%)
	87	6.2±3.9			8 (9.8%)		3 (3.7%)		87*	-	7(8%)
Ovrouski S	15	26(16-38)	6.2	10 (71%)	8 (57%)				10* 5**	1 (6.6%)	1(6.6%)

* - intra-atrial lateral tunnel technique; ** - extracardiac conduit technique; AP - atriopulmonary; CP - cavopulmonary; PLE - protein-losing enteropathy; HF - heart failure; Imm - immediate; m - months; TE - thromboembolism; AP technique may contain rare cases of the atrioventricular technique.

Table 1 - Follow-up complications of the Fontan operation, according to several authors

considerably lower (60%) than that of 94% reported by Nakano et al¹⁴ in 167 patients undergoing surgery from 1991 to 1999, mean age of 6.2 years, after a 10 and 8-year postoperative follow-up, respectively.

That is why a standardization characterizing the need of an earlier intervention is questionable, because it could sometimes interrupt the natural course of patients whose oxygen saturation remains high and adequate for a long time, until the intervention becomes necessary due to the onset of hypoxia.

At Instituto do Coração da Faculdade de Medicina da Universidade de São Paulo, the standardization of an early indication has not been made in most of the situations, and in 41 patients undergoing cavopulmonary connection with atrial fenestration, mean age of 7.6±4.6 years, and in other 21 additional patients without atrial fenestration, mean age of 7.3±3.4 years, the outcome has proven adequate (57 out of the 62 patients in functional class I) in a mean follow-up period of 3.1±2.4 and 5.1±3.3 years, respectively, in the two groups¹⁵.

It is worth pointing out that Binotto et al¹⁶ found interstitial fibrosis mainly in the subendocardial layer in necropsy studies, in inflow sites, in the left ventricular apex and outflow tract in tricuspid atresia, and in fibrosis extension proportional to the patient's age. This finding represents a chronic volume overload in the presence of ischemia occurring early in life, since the age of this group studied corresponded to 9.6±13.9 months, a mean of 2.5 months.

Despite the demonstration of fibrosis increasing with age in

patients with heart diseases such as tricuspid atresia, it remains to be demonstrated whether this would occur if the patient had a steady course, with balanced pulmonary and systemic flow, until the surgical intervention became really necessary in view of the natural progression of hypoxia.

The favorable outcome demonstrated by patients undergoing surgery even in adulthood¹⁰ is in favor of this proposition.

Much remains to be proven in this respect, but it is necessary to always bear in mind the Fontan operation is a palliative intervention and has a series of inconveniences such as the complications already mentioned, as well as the demonstration of elevation of α -1-antitrypsin even in the absence of hipoproteinemia¹⁷, the subclinical coagulation disorders¹⁸, and the elevation of systemic venous pressure itself, that place the outcome of these patients at risk.

The preservation of all the adequate factors for a successful surgery indicated early or later in life does not imply a perennial normal cardiac function, given the anatomic and functional change imposed by the total cavopulmonary technique which, in itself, predisposes toward the unconditional occurrence of the aspects related to the complications mentioned.

However, we remain optimistic about this technique, given that the substantial change induced by the restoration of normal arterial oxygen saturation is clearly superior in relation to that provided by other palliative techniques which, on the contrary, lead to cardiac overload, as is the case of the classic systemic-pulmonary shunt (Blalock-Taussig procedure). Additionally, the cavopulmonary operation is, in thesis, a

technique that eliminates heart overload because the venae cavae and the pulmonary arterial tree are placed in series.

For all these reasons, today it seems evident the Fontan operation may bring an even more favorable outcome, provided that the acquired factors are minimized with earlier corrections, as from two years of age, and the technique chosen is that of the use of an extracardiac conduit, in order to obtain less arrhythmias, congestive phenomena, and protein-losing enteropathy, thanks to the elimination of intra-atrial suture lines, of damage to the sinus node, and reduction in factors of increased atrial pressure and volume, these latter ones being favored by the intra-atrial lateral tunnel.

I take this opportunity to mention, still as only perspectives, other advantages of the cavopulmonary operation with extracardiac conduit over the intra-atrial lateral tunnel that should be pointed out. Among the major advantages is the applicability of the technique in patients with heterotaxia and with alterations in systemic and pulmonary venous return, as well as with an atrioventricular valve, the rare occurrence of supraventricular arrhythmias, with the possibility of concluding the cavopulmonary connection without the need of a cardioplegic cardiac arrest, in addition to hemodynamic advantages that do not predispose to systemic venous congestion⁸.

Atrial fenestration is not mandatory, provided that the factors indicating the technique are preserved and later corrections are not ruled out, mainly in adulthood, provided

that the elements of a good indication are preserved. We also broaden the concept that fenestration should not be a fundamental procedure, provided that the decrease in pulmonary pressure is obtained with other therapeutic methods in the immediate postoperative period, given the availability of the use of inhaled nitric oxide and of other vasodilating agents, in addition to early anticongestive measures that lead to a reduction of edema in general.

Furthermore, the need of a long-term anticoagulation seems to be a consensus in order to minimize the occurrence of thromboembolism.

In the immediate postoperative period, aiming at reducing the risks and the intensity of pleural effusion, as well as the duration of hospital stay, Cava et al¹⁹ suggest the use of intravenous furosemide at a 1mg/kg dose every 8 hours in the first day, and, after 2 or 3 days, the combined use of hydrochlorothiazide and spironolactone at a 1 mg/kg dose every 12 hours, in addition to captopril 1mg/kg daily, and severe water restriction.

This presentation can be summarized by pointing out the favorable outcomes of the total cavopulmonary operation without fenestration attributed to the beneficial effects of the extracardiac conduit and to the good selection of patients.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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