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Review

Converting the Fontan Circulation: Challenges and Evolution

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Abstract

The Fontan operation, the final palliative step after a series of complex operations in patients with univentricular hearts, has undergone multiple modifications throughout the last decades, with the goal of finding the method which combines the optimal hemodynamic effects of the Fontan circulation with minimal long term side effects. An understanding of the operative evolution and subsequent side effects, as well as the management thereof seems imperative. Since its inception by Francis Fontan the, now obsolete, initial atriopulmonary connection has passed through several milestones into having now reached the era of total cavopulmonary connection. However, recently published results bring to light the new option of biventricular conversion which comes to challenge the management of Fontan patients as we know it. Currently, there is no consensus as to whether to continue with palliation in this challenging group of patients or proceed with heart transplantation. In this collective review, we provide a historic overview of the Fontan evolution as well as future insights, discussing the perspectives of options for patients with failing Fontan, including the latest addition of biventricular correction.

Keywords

Fontan circulation; Fontan conversion; total cavopulmonary connection; single ventricle physiology

Introduction

First described by the pioneer Francis Fontan in 1971 [1], the Fontan operation has enabled thousands of children with congenital heart disease to survive and reach adulthood. The original technique is known as the atriopulmonary Fontan connection and many variations that followed were based on the principle that the right atrium can act as the pump for the pulmonary circulation. Subsequently, de Leval [2] thoroughly questioned the role of the right atrium, and in the contemporary design of the Fontan operation, the superior vena cava is anastomosed directly to the pulmonary artery and, using a conduit that is created inside the baffle, the inferior vena cava is also drained to the pulmonary artery (lateral tunnel technique). This technique comes with many advantages, such as the decreased risk of thrombosis due to less blood stasis, as well as the exposure of only a limited portion of the right atrium to high venous pressures, thus lessening the risk of arrhythmias. In addition, the coronary sinus remains in the low-pressure atrium so that the myocardial venous drainage is left unharmed. The most recent modification of the Fontan operation technique includes the use of an extracardiac interposition graft between the transected inferior vena cava and pulmonary artery [2,3].

Nowadays, the two aforementioned techniques of total cavopulmonary anastomosis have been established as operations of choice, and the initial technique of atriopulmonary connection has been abandoned.

This article aims to review the current experience and bibliography, strong points and limitations, regarding Fontan conversion, 25 years after its conception. Moreover, the evolution and latest surgical techniques regarding the failing Fontan circulation will be studied.

Origins of Fontan Circulation

Historical Background

To minimize Fontan failure drawbacks and postpone heart transplantation, a pioneering surgical conversion to total cavopulmonary connection was introduced by Mavroudis in patients with an atriopulmonary connection and preserved ventricular function [4]. Because of the complications that arose, many patients who had undergone an operation with the atriopulmonary anastomosis technique were led to conversion operations for total cavopulmonary anastomosis. These complications include obstructions in the venopulmonary artery pathway, pulmonary arteriove-

nous malformations, atrioventricular valve regurgitation, and atrial arrhythmias, among others. The extremely important role of atrial tachyarrhythmias in patient survival led to not only converting the atriopulmonary Fontan to a total cavopulmonary connection but to include atrial arrhythmia surgery and pacemaker implantation in the whole process, thus creating the Fontan conversion surgery, with excellent results [5,6].

However, even with those additions, the Fontan circulation in patients with converted Fontan or younger ones with total cavopulmonary connection is deemed to fail in the long run. Taking into consideration the small number of available organs for transplantation and the relatively worse outcomes compared to other congenital heart conditions, new surgical techniques were needed in order to further "convert" the Fontan physiology without the need for a heart transplant [7,8]. The newest established technique is the Biventricular conversion in patients with failing Fontan circulation, though adequate ventricular size and function are needed [9,10].

Failing Fontan Physiology

Fontan failure is a loosely applied term with no clear distinction regarding its timeline. It is safe to assume though, that from the very start this circulation is inherently inadequate in comparison to the population with no congenital heart disease regardless of the excellent short-term and early surgical outcomes [11]. During the early stages, Fontan failure is characterized by exercise intolerance and an increase in New York Heart Association (NYHA) class status and quality of life. In later stages, there are more severe signs whose incidence is markedly increased in atriopulmonary Fontan, like ascites, protein-losing enteropathy and plastic bronchitis. The majority of these complications are universal in the earlier era and the latest surgical techniques, however, they tended to be more common and present sooner in the historical atriopulmonary Fontan. This is caused by the gradual atrial dilatation, leading to increased hepatic pressure, reduced pulmonary artery blood flow and compression of pulmonary venous return which can lead to pulmonary vein obstruction, while the atrial scarring as a result of hypertrophy and dilatation leads to atrial reentrant tachyarrhythmias. This leads into a vicious cycle because these tachyarrhythmias further worsen atrial function, atrioventricular synchrony and cardiac output. Both the hemodynamic and electrical complications of atriopulmonary Fontan can lead to thrombus formation and systemic vein obstruction. This kind of atrial dysfunction is of substantial importance considering the passive venous circulation in Fontan patients. The loss of the atrial structural and functional architecture is one of the reasons that total cavopulmonary anastomosis is the treatment of choice for Fontan patients and also why the Fontan conversion procedure was introduced in the first place [12–15].

Evolution and Research Advancements of Fontan

Fontan Conversion

As previously mentioned, the Fontan conversion procedure was first introduced in 1998 by Mavroudis *et al.* [4]. This procedure consists of three major parts which include (a) revision of the atriopulmonary to extracardiac Fontan, (b) atrial arrhythmia surgery and (c) pacemaker implantation. In its early stages, Fontan revision for hemodynamic indications was thought to reduce atrial arrhythmias by improving hemodynamics and atrial wall strain, but the arrhythmic load was not eliminated and especially early postoperative atrial arrhythmias severely compromised atrial output, thus increasing mortality [16,17]. A retrospective comparison between Fontan revision and Fontan conversion showed improved outcomes in the latter category [18]. Although the need for a pacemaker is not always highlighted, it allows the patient to avoid resternotomy, especially considering the possibility of atrioventricular conduction abnormality and the need for advanced antiarrhythmic medication or atrial pacing in atrial arrhythmia recurrence. It is also important to note that by the end of the Fontan conversion procedure the only blood flowing through the right atrium is that of the coronary sinus, passing freely through the atrial septal defect. This drastically reduces the transcoronary pressure improving heart blood supply [19].

Fontan Conversion Experience in the Last 25 Years

With the progression of the Fontan conversion procedure and the ever-increasing experience of congenital cardiac surgeons, gradually more complex procedures took place for comorbidities regarding Fontan failure. Valve repair or replacement, atrial resection, and pulmonary artery reconstruction were added to the procedure, while advanced antiarrhythmic surgery and lesion-specific cardio ablation took place in order to fully treat atrial arrhythmias. When performed by an experienced multidisciplinary team, with careful patient selection criteria applied, there was no increase in mortality or morbidity [5,20–23].

Moreover, the increased experience allowed for earlier interventions in atriopulmonary Fontan patients, in order to proactively prevent the associated manifestations of a failing Fontan and increase transplant-free survival. Mavroudis and colleagues [19] studied 149 cases of Fontan conversion showcasing some interesting results which can be considered intuitively obvious.

Diller *et al.* [14] reviewed 321 adult Fontan patients in a multicenter study and proved that the presence of atrial arrhythmias led to a 3-year mortality of 25% and a 6-year mortality greater than 40%, while no arrhythmias led to a 6-year mortality of around 10%. This confirms the signifi-

cance of the antiarrhythmic component of the Fontan conversion procedure [14].

The Australian and New Zealand registry of Poh *et al.* [24], studying patients undergoing Fontan conversion between 1990 and 2014, showed that early conversion centers had significantly improved transplant-free survival compared to centers where the procedure was carried out after late Fontan failure [24].

In the paper by Mavroudis *et al.* [19] 149 cases review, there was a 2% perioperative mortality, with an excellent 10-year 84% freedom from death or heart transplantation. There were also 12 patients who received heart transplantation, half of which were within one year, thus considered conversion failures. At 15 years post conversion, freedom from arrhythmia was found to be 80%. This is partly due to the intraoperative atrial lesion cryoablation, and partly due to the atrial pacemaker providing atrioventricular synchrony and reducing premature atrial contractions and atrial reentry tachycardia [19]. This has been further proven by Terada *et al.* [25] in a 25 consecutive patient study where there was concomitant pacemaker implantation in six of of the patients at the time of conversion. The results showed that five patients required postoperative pacemaker implantation that could have been avoided if there was an implantation at the time of surgery. The authors concluded that pacemaker implantation is necessary in all Fontan conversions which include right or biatrial maze procedure [25].

The success of this series is not only based on the surgical expertise and significant experience of the teams that carried them out, but also on careful patient selection. Patients with severely impaired right atrial function or patients with new-onset arrhythmia who preserved a NYHA class II status, responded excellent to Fontan conversion [19,24,26]. At this point, it is worth mentioning that there are many limitations in transcatheter ablation of atrial arrhythmias due to the thickness of the right atrium. This is supported by small series (6 patients were included) from Betts *et al.* [27] where most of the Fontan patients who underwent ablation of atrial arrhythmias had a recurrence and have undergone Fontan conversion. There are, however, studies such the study by Correa *et al.* [28] with a population of 118 patients who have undergone Mustard, Senning or Fontan operations and have developed atrial tachycardias, suggesting that transcatheter ablation is feasible with complete arrhythmia treatment of 58%, though it is pointed out that the outcomes are worse in Fontan patients.

Current Challenges

Fontan Conversion or Heart Transplant?

Patients that have undergone atrioventricular Fontan are an extremely fragile population. It is important to note that Fontan failure is an evolving condition, and these patients are constantly deteriorating in terms of NYHA class, multiorgan failure and cardiac function. Thus, the most important factor for the surgeon and for the patient is timing [24,29,30]. In this particular case, that is due to the very difficult question of whether to convert the Fontan and continue palliation or to proceed with heart transplantation (Fig. 1) [31,32].

There are several factors favoring either one or the other. Fontan conversion is generally preferred in patients with a preserved ventricular function and a severely dilated right atrium or patients with a recent decrease in ventricular function related to new onset atrial arrhythmias. In addition, patients who have not yet had a bilateral Glenn as part of their total cavopulmonary conversion have improved outcomes after Fontan conversion. Regarding new-onset arrhythmias, the Down Under guidelines by Dr. Poh and colleagues [24], studying a population of 39 patients from six centers in New Zealand and Australia, recommend that patients should be referred within three years of their arrhythmia and while still in functional NYHA class II. They should also be referred before starting a second antiarrhythmic medication and as previously stated due to the dramatic thickness of the right atrium, transcatheter ablation is not recommended [24,29,33].

It has also been observed by Mavroudis *et al.* [19], that some patients undergoing Fontan conversion had cardiac death or a need for heart transplantation soon after surgery. The risk factors regarding that are a right or indeterminate ventricular morphology, a need for a biatrial maze in complex arrhythmias, a cardiopulmonary bypass time >240 minutes and preoperative ascites or protein-losing enteropathy.

On the other hand, there are occasions when it is too late and there is no benefit in Fontan conversion. Such patients are referred for heart transplantation. The factors towards heart transplantation are end-stage ventricular failure, arrhythmias for more than five to ten years, plastic bronchitis, protein-losing enteropathy, ascites and moderate or worse aortic valve regurgitation [30,34]. It is considered that the abovementioned conditions cannot be reversed by Fontan conversion. Another interesting observation is that patients who have undergone Fontan conversion did not have an increased mortality after heart transplantation compared to patients without conversion [8,19,35,36]. Moreover, as studied by Riggs *et al.* [37], it is worth noting that patients who have undergone the Fontan operation are in no way inferior in terms of survival after heart transplantation compared to transplant recipients with a congenital heart disease and biventricular physiology, if at least a year has passed since the Fontan operation.

There have also been studies considering the use of ventricular assist devices in failing Fontan population. These studies mostly consider the ventricular assist device implantation in selected patients as a bridge to transplant

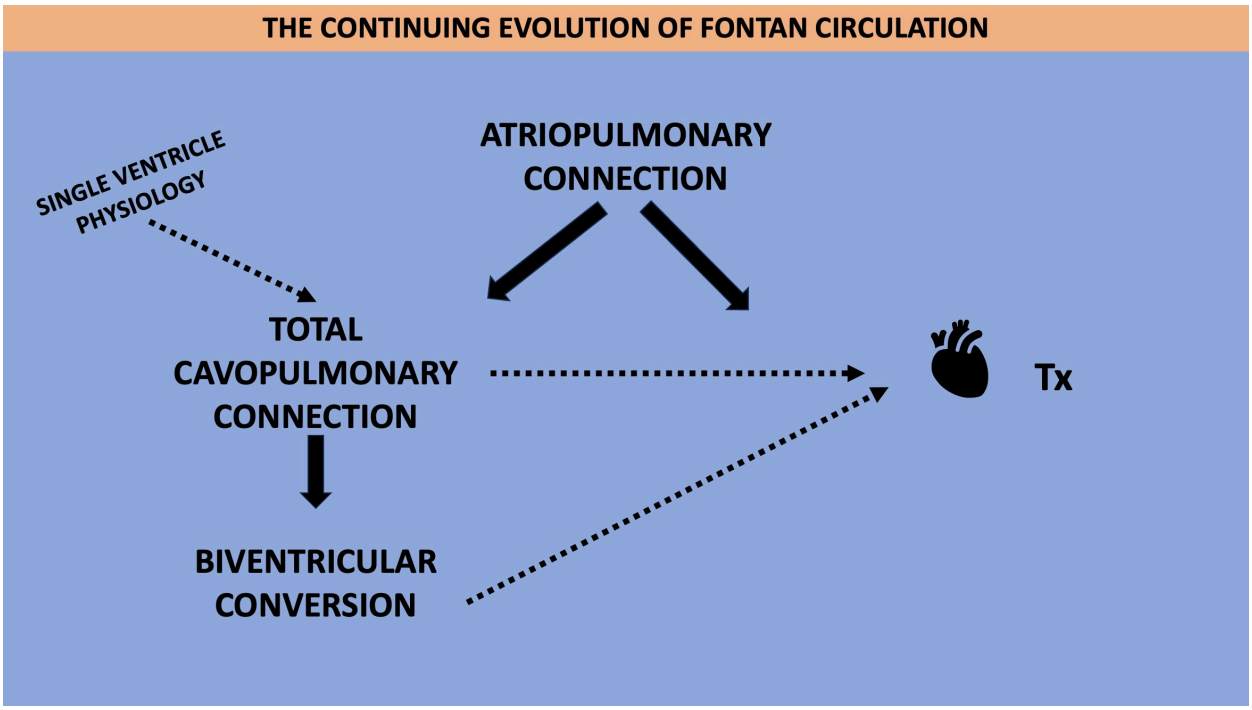

Fig. 1. The continuing evolution of Fontan circulation. Management options following the now obsolete atriopulmonary connection. Tx, Transplantation.

with promising results [30,38]. Since its first successful use in 2005 many steps have been made both surgically as well as mechanically in ventricular assist devices [39]. Since then, there have been some successful cases with good results but there have been no big studies concerning their use in failing Fontan circulation [40,41]. That was until the last three years where a retrospective study by Cedars *et al*. [42] in 45 Fontan patients with ventricular assist device (VAD) implantation was released. It showed a 1-year survival of 78.7% with 69.5% rate of successful transplantation and 9.2% still on ventricular support at the time of the study. This study was conducted through the use of Advanced Cardiac Therapies Improving Outcomes Network [42].

What is Next in Failing Fontan Circulation?

The patient pool of atriopulmonary Fontan is ever-reducing due to newer and better techniques, cardiac transplantation, conversion, or death. The current Fontan population consists of patients who have either undergone total cavopulmonary connection or converted their atriopulmonary Fontan. This patient population ages and increases and their circulation is gradually failing so it is of utmost importance for a new surgical solution to be found [26,29,31].

The best and most definitive solution is heart transplantation but there can never be enough available organs for transplanting all these patients and the waiting list times are continually growing. Heart transplantation in these cases has worse outcomes than congenital heart disease generally. This is in part due to the aforementioned long waiting lists and late referrals, but mostly due to multiple previous operations, elevated panel reactive antibodies, hepatic and renal dysfunction and significantly increased surgical complexity, since this is not a straightforward cardiac transplantation and it requires concomitant operations such as pulmonary artery reconstruction and several others as stated by Jayakumar *et al*. [43].

Moreover, there are adequately studied long-term complications associated with Fontan circulation, affecting patients' quality of life and leading to adverse events. These consist of atrioventricular regurgitation or stenosis, pulmonary or systemic venous obstruction, presence of venovenous or arteriovenous collaterals, left ventricular outflow tract obstruction, protein-losing enteropathy and plastic bronchitis. Each of these has its own surgical or interventional treatment with varying degrees of success, but most of these cases are extremely challenging with no definitive solution [19,26,29].

Biventricular Conversion: New Horizons in Fontan Physiology

Having that in mind, the technique of biventricular conversion (BiVC) was proposed. BiVC consists of Fontan circulation takedown and redirecting the blood so as to flow physiologically from the left ventricle to the aorta and from the right ventricle to the pulmonary arteries using atrial and ventricular septation. The timing of this procedure primarily depends on the ventricular size. When it is adequate,

Fontan takedown and BiVC can be done in a single procedure, which is termed as primary BiVC. When the size of the ventricle is inadequate, the surgical team can employ recruitment strategies, which refer to concomitant surgeries such as atrial septal fenestration creation, ventricular septal defect closure, valve correction, left ventricular outflow tract obstruction resection *etc*. and lead to staged recruitment of the nondominant ventricle. These patients undergo Fontan takedown and BiVC at a later stage which is referred to as staged BiVC [10].

A single center experience of 23 patients who have undergone Fontan takedown and BiVC by Doulamis *et al.* [9] shows promising results though more studies are needed. Specifically, primary BiVC was found to be feasible in patients with mild hypoplasia and staged BiVC was preferred following ventricular recruitment in moderate to severe left heart hypoplasia. The results were found to be better in well-selected patients with failing Fontan circulation and especially good when there was an elective takedown without failure indications, while they were worse in patients with severe Fontan complications. The mortality in the group that underwent selective takedown and BiVC was 0% while in the failing Fontan subgroup it was 33%. This raises the question of the proper timing for biventricular conversion and whether it should be used in patients with a stable Fontan circulation considering the long-term adverse outcomes of the Fontan circulation, which were previously described [9].

Another study by Kumar *et al.* [10] states that biventricular conversion is a viable strategy at every point of a single ventricle physiology patient's life but only if ventricular size and function allow it. Moreover, it is a viable option in patients unable to establish Fontan circulation due to unfavorable pulmonary artery anatomy, elevated pulmonary resistance, or complicated systemic venous anatomy. It is also indicated in patients who have developed severe extracardiac complications [10].

The cornerstone in patient selection and biventricular conversion planning is a multidisciplinary approach by an experienced heart team, as well as multimodality imaging with all the available methods in order to specify the anatomy as well as the function of all the cardiac and vessel components. From the limited thus far experience there are factors derived from magnetic resonance, computed tomography, right heart catheterization, and ultrasound associated with a successful biventricular conversion. At this point it is imperative to highlight cardiac magnetic resonance as a means to distinguish the proper intervention timing by calculating cardiac fibrosis. Also, three-dimensional printing, especially in such complex cases, is of paramount importance and provides essential information for venous inflow and atrial anatomy, atrioventricular valve morphology and outflow tract anatomy in order for the operation to be successful [9,44,45].

Future Perspectives

There has been great improvement in management and long-term outcomes concerning single ventricle physiology and the relevant surgical treatment. This is largely due to increasing medical expertise and modern surgical techniques as well as more available resources. The fact is though, that the Fontan procedure and its conversion are decreasingly relevant, even though these are historic and monumental procedures. Total cavopulmonary conversion is a well-established surgical treatment, though it still does not prevent many extracardiac complications of single ventricle physiology. As these patients get older there needs to be a next step in their treatment before heart transplantation primarily due to the small availability of heart transplants. It is also worth noting that these patients who have undergone multiple operations, have had continued exposure to elevated intracardiac end-diastolic pressure and increased exposure to homograft usage might not be the best candidates for transplant, so the management needs to steer from heart transplantation and be used as a last resort in cases that allow it.

Dr. de Leval stated that Fontan's operation challenged the landmark 1628 physiology study of William Harvey regarding the human circulation. Similarly, the biventricular conversion now challenges the norm and routine practice of surgical management in Fontan circulation patients.

Biventricular conversion seems to be a possible solution to the abovementioned and several reports have established its feasibility in children with complex heart defects and borderline anatomy. What remains to be seen though, is the long-term outcomes, since there are no relevant large studies. Results appear promising in the reversal of some of the Fontan circulation complications as protein-losing enteropathy and plastic bronchitis. In contrast, it is imperative to highlight the high need for reoperations in BiVC, either surgical or interventional for residual lesions that severely impact patients' quality of life. This is another factor that needs to be reconsidered in these cases since not all patients can tolerate multiple reinterventions. At this point, since there are inadequate studies, whether a BiVC or a univentricular repair is better remains to be seen. There is no realistic control group and since the long-term complications are our area of interest concerning these patients the real benefit may be realized after many years [9,10,44,45].

Conclusion

Fontan conversion is a well-established surgical procedure that increases survival in patients with single ventricle physiology. In patients with total cavopulmonary connection or with converted Fontan, new options are needed in order to increase survival. Biventricular conversion seems

to be such an option, with good short-term results, especially with optimal timing in selected patients. A multidisciplinary approach by the heart team, with multimodality imaging provides the most useful amount of information to plan these procedures and select the patients which would benefit the most, as well as the optimal timing. Heart transplantation remains an option in carefully selected the patients in order to have the desired result. Due to the limitations of such patients, concerning sparsity and complexity, a multi-institutional, multinational collaboration seems necessary so as to properly study these patients and offer them a tailored approach with the best possible results.

Author Contributions

MKa, AC, and FPR designed the research study. MKa, AC, and FPR performed the research. TC, MKo and DB analyzed the data. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

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Conflict of Interest

The authors declare no conflict of interest.

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