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Fontan Circulation: How a Model can Hinder or Help to Understand and Manage a Complex Problem

Marc Gewillig[1*], Thomas Salaets[1], Alexander Van de Bruaene[1], Stephen C Brown[1,2]

Abstract

When faced with complex circulatory issues, a model can assist the clinician in understanding, manipulating, and treating the underlying problem. The creation of a Fontan circuit currently consists of connecting the caval veins to the pulmonary arteries without the interposition of a pumping chamber. Complications of this univentricular circulation carry a significant morbidity and mortality and are notoriously difficult to manage. The most obvious and intuitive model places the pulmonary vasculature downstream of the ventricle, impeding the run-off after the systemic capillaries. This implies that the ventricle can overcome the downstream problems, which in clinical practice has been shown to be incorrect. Another possible but less intuitive model for the Fontan circulation is putting the pulmonary circulation upstream of the ventricle, thereby restricting the preload. Here the pulmonary circulation acts like a dam creating upstream congestion and downstream decreased flow inertia. This model allows much better to predict the observed changes in the cardiovascular system over time. Based on this concept, the late ventricular deterioration will not be the result of excessive ventricular afterload but secondary to chronic volume depletion/underloading. A model can be good at one level but inadequate or even incorrect at another level; the choice of the model may hinder or help the understanding of the problem.

Keywords: Fontan; Preload deprivation; Afterload; Failure; Bottleneck

Introduction

When confronted with complex problems or unfamiliar situations, our brains prefer to work with a model: this facilitates understanding and allows predictions of how the circuit may react to interventions or evolve over time. Interventions based on a rational model facilitate efficient targeted manipulation or therapy.

The Fontan circulation is apparently simple, but its function and controls are poorly understood and associated with multiple problems that are hard to avoid or cure. A "Fontan circulation" currently consists of connecting the caval veins to the pulmonary arteries without the interposition of a pumping chamber [1] (Figure 1 A-C). In this construction, residual potential and kinetic energy of the systemic venous blood are used to push blood through the lungs in a new portal circulation-like system [2]. Complications of this circulation include early and late mortality, exercise intolerance, ventricular dysfunction, rhythm and conduction disturbances, hepatomegaly with secondary fibrosis, cirrhosis and carcinoma, systemic venous thrombi, lymphatic dysfunction with protein-losing enteropathy and bronchial casts, ascites, pleural effusions and peripheral edema [3,4].

Affiliation:

[1]Pediatric & Congenital Cardiology, University Hospitals Leuven, and the Department of Cardiovascular Sciences, Catholic University Leuven, Belgium

[2]Pediatric Cardiology, University of the Free State, South Africa

***Corresponding author:**

Marc Gewillig, Department of pediatric and congenital cardiology, University Hospitals Leuven, Herestraat 49, B-3000 Leuven, Belgium

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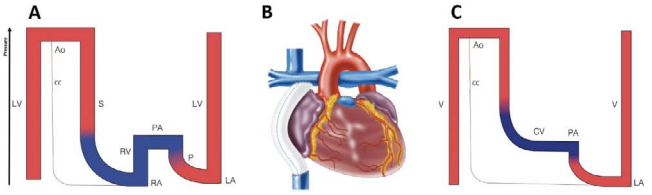
After the creation of the Fontan circuit, secondary and tertiary changes occur in this circulation at multiple levels, including at the level of the single ventricle. The ventricle typically has a cavity bigger and a wall thicker than expected for body surface area (BSA), overall hypocontractility, and filling pressures which initially can be good but invariably deteriorate over time! [2]. This eventually may lead to severe secondary systolic and diastolic dysfunction. A model is warranted to better understand these phenomena, which is crucial for targeted, efficient interventions.



A: schematic representation of a normal circulation: the pulmonary circulation (P) is connected in series with the systemic circulation (S). The right ventricle maintains a right atrial pressure lower than the left, providing enough energy for the blood to overcome the pulmonary resistance**. B & C: cartoon and schematic representation of a TCPC Fontan circuit:** the caval veins have been connected end-to-side to the pulmonary artery. The post-capillary energy is collected in the caval veins and used for trans-pulmonary flow. Ao: aorta; CC: coronary circulation; CV: caval veins; LA: left atrium; LV: left ventricle; PA: Pulmonary artery; RV: right ventricle; V: single ventricle. Line thickness reflects output, color reflects oxygen saturation.

Figure 1A-C: Schematic representation of a normal (A) and a Fontan circulation (B-C)

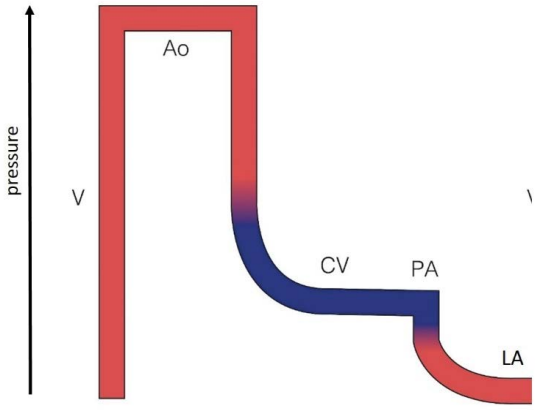
Intuitive model – impeded run-off

The most obvious and intuitive model places the pulmonary vasculature downstream of the ventricle, impeding the run-off (Figure 2). It can be derived from the diagram in Figure 1 and explains where the energy is derived from to ensure flow through the lungs.

Based on this model, the clinician will try to determine the new controls in the circuit. The systemic and pulmonary resistances are put in series and, therefore, as in an electrical circuit, are frequently added up, suggesting that the workload to the ventricle has increased. This model can further be simplified, as in electrical models, to a single resistance at the exit of the single ventricle, akin to putting a clamp on the aorta causing a moderate obstruction. Many clinicians dealing with patients with a Fontan circulation have used this model for decades- knowingly or unknowingly.

This "impeded run-off" model predicts that the ventricle will eject against a higher resistance since the afterload to the ventricle is increased following the creation of the Fontan circuit; the cavity may, therefore, acutely enlarge and wall thickness may increase in time. Furthermore, the contractility may deteriorate, and filling pressures may rise as the hemodynamic situation deteriorates. As mentioned, clinicians see many of these features occur while following patients with a Fontan circuit.

Moreover, this model suggests that the ventricle can be used to overcome the downstream hemodynamic problems: in cardiology, clinicians frequently encounter situations with increased afterload. This model predicts that inotropes should be helpful, as should be systemic vasodilators; tuning the heart rate might also improve the hemodynamic burden.



The pulmonary circulation is put downstream of the ventricle impeding run-off after the systemic capillaries. Symbols as in figure 1.

Figure 2: Fontan cardiovascular model "impeded run-off".

With this model in mind, clinicians have been trying to find evidence of its robustness and, last but not least, find efficient, targeted solutions for compromised Fontan patients with a progressively declining circulation. However, in the real world, the results of this search were very disappointing. Several studies have been conducted to prove that inotropes, systemic vasodilators, or heart rate manipulation could improve output or congestion; so far, no single controlled study has shown conclusive evidence for this [5-7]. Studies were then repeated with larger numbers of patients, with other and more sophisticated variables or compound endpoints, but still, no study was able to show significant improvement [8].

On the other hand, critical analysis of the model reveals that significant deviations from reality can be observed. Immediately after creating a Fontan circuit, the presumed acute increase of afterload should result in an increase of ventricular volume with thinning of the walls, later followed by hypertrophy. However, careful clinical observation following Fontan operation demonstrated that the ventricular cavity shrinks from baseline but frequently remains bigger than expected for BSA. Also, the wall immediately after the operation does not show a phase of initial thinning before thickening as suggested by this model, but immediately thickens as the ventricular muscular mass is redistributed

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around a smaller volume [9]! Moreover, while the model suggests that the afterload should increase, simple reasoning and common sense suggest the opposite! Afterload to the ventricle reflects the force needed by the ventricle to eject a specific volume against a certain pressure into the aorta; in a Fontan circulation, these 2 exclusive variables of afterload– pressure and volume- invariably decrease, making the suggestion of an increased afterload sound unscientific [10].

The term afterload is often used loosely by physiologists and clinicians; afterload can be related to the myofiber, the sarcomere, the myocardial cell, to the myocardial wall, or to the ventricle as a pump. It can be defined in its most basic physiological way as all factors contributing to total myocardial wall stress (or tension) during systolic ejection [11,12]. When related to the wall, afterload is best described by LaPlace's laws, which include the major determinants of end-systolic ventricular pressure and radius, as well as myocardial wall thickness. However, multiple other factors, e.g., outflow tract resistance, arterial pressure, arterial wall compliance, blood volume, and peripheral resistance, are all additional factors that also determine afterload [1].

The possibility that a model has flaws or is inappropriate is frequently challenging to acknowledge for many and various reasons: desire to make the model fit, lack of alternative model, scientific or financial bias, etc. Such obstinacy is not uncommon in human history. For centuries, Aristoteles, Ptolemaeus, and the Catholic Church promoted – each for their own reasons- the geocentric model, whereby the earth was considered the center of the universe, with all (then known) planets, including the sun, revolving around it. The geocentric model is easy to understand by an individual who observes the sun rising in the east and setting in the west. The geocentric model allows the prediction of simple local events, e.g., how a shadow will advance on Earth. However, the geocentric model appeared insufficient when more complex activities were contemplated, such as launching a satellite to another planet. Despite fanatic and bloody repression for centuries, the geocentric model has finally been overshadowed by the heliocentric model, but only many decades after it was first described by Copernicus [13].

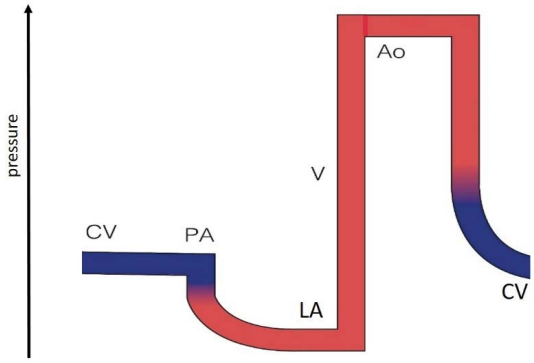
Alternative model – restricted preload

Another possible but less intuitive model for the Fontan circulation is putting the pulmonary circulation upstream of the ventricle, thereby restricting the preload. This model is based on the concept of constructing a dam wall in a river to regulate water supply downstream. Like a dam, the resistance of the pulmonary vascular bed will create upstream congestion and decrease the blunted inertial flow downstream, which in a closed circuit will result in overall decreased output [2]. Thus, by creating a Fontan circuit, this "dam wall" (i.e. pulmonary neo-portal system) becomes the new critical bottleneck: it will determine the flow throughout the circuit (Figure 3) [14].

The pulmonary circulation is put upstream of the ventricle, causing upstream congestion and downstream decreased flow. Symbols as in figure 1.

Applying this model, the following early changes can be predicted: pressure in the caval veins will rise, while all pressures and flows downstream (left atrium, single ventricle and aorta) will decrease; the left ventricular cavity will decrease from pre-Fontan status [which is larger than expected for BSA due to the fetal univentricular circulation and the volume load caused by the initial palliation(s)] with an immediate proportional increase of wall thickness [3]. Contractility indices will decrease (Frank-Starling mechanism), diastolic pressure will (initially) decrease, and systemic vascular resistance will increase to maintain perfusion in critical organs as aortic pressure decreases. Consequently, the ventricle's afterload (which is very preload dependent) will decrease as pressure and stroke volume decrease.

Applying this model to the Fontan circulation,



The pulmonary circulation is put upstream of the ventricle, causing upstream congestion and downstream decreased flow. Symbols as in figure 1.

Figure 3: Fontan cardiovascular model "restricted preload"

recommendations for targeted early treatment can be suggested. Augmenting systemic venous pressure will increase overall flow but, unfortunately, also congestion; lowering the resistance over the Fontan portal system will improve flow, as will reducing the left atrial pressure. Other strategies, such as increasing systolic function, lowering systemic vascular resistance, or tuning heart rate, are unlikely to have an effect unless they lower (in time) the left atrial pressure. Changes in the ventricle are not due to an intrinsic myocardial problem. These can be attributed to the markedly different loading conditions created by the pulmonary vascular "dam wall": severe chronic volume deprivation. If the damming is excessive, this will result in excessive congestion, edema and effusions, low overall output with hypotension, and eventually death. This is precisely what the clinician observes early after the creation of a Fontan

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circulation! Although this is not proof of the adequacy of the model, it assists the clinician in understanding the changes and allows appropriate manipulation of the Fontan circulation.

The model also allows one to predict changes in the cardiovascular system over time. Based on this concept, these will not be the result of ventricular overload but secondary to chronic volume depletion or unloading.

Chronic volume deprivation, either by disease or lack of exercise or a sedentary lifestyle, is known to cause a ventricle to become less compliant with increasing filling pressures [15-17]. Thus, initially, the left atrial pressure will decrease at the time of the Fontan operation, but over time, it will increase due to chronic volume deprivation and reduced or absent exercise-induced ventricular stretch. However, this rise in left atrial pressure has an enormous deleterious effect: flow through the critical bottleneck (pulmonary bed) is further hindered by poor runoff into the ventricle, resulting in increased retrograde congestion and impaired cardiac output, further depriving the single ventricle. Reduced preload will further reduce systolic function (Frank-Starling mechanism), and eventually, when the ventricle is severely deprived and reaches its closing volume, systolic function will disproportionately worsen [18]. At this point, the "limited preload" model reaches its limits, and more conventional cardiac models typically dealing with "primary" myocardial dysfunction also come into play.

The "impeded runoff" model can still be used, but resistances should not simply be put in series such as for an electrical model, but rather like a waterfall cascade: upstream of the cascade, at the level of the source (in casus the aorta) one does not see what happens downstream beyond the waterfall (unless severe congestion occurs). Adding resistances further downstream from the waterfall has no effect upstream of the cascade unless overall flow is diminished, as in this closed circuit. The "impeded runoff" model is suitable for envisioning where energy comes from for transpulmonary flow; the "restricted preload" model better explains flow restriction and allows improved understanding and predictions early in the Fontan circuit.

Young individuals will start understanding the sun's motion with a geocentric model but must later be guided to the heliocentric model. Similarly, new generations of physicians will intuitively begin with the "impeded runoff" model but should be guided by their mentors to the "restricted preload" model. This model however also lacks power when the systolic function decreases disproportionately to the decreased preload and the ventricle reaches its closing volume or suffers from previous/additional damage. As always, any model should be used cautiously and be examined critically as they all have limitations, especially when the problems they are supposed to explain become more complex.

Conclusions

The human brain develops models to understand problems and make predictions allowing targeted interventions. Such a model can be good at one level but inadequate or even incorrect at another level or time period; the choice of the model may hinder or help the understanding of the problem. Applying the most likely model to the Fontan circuit, it is evident that the upstream neo-pulmonary portal system becomes the main determinant of flow early after the establishment of a Fontan circuit. Late additional ventricular dysfunction is as such not the result of intrinsic myocardial abnormalities or systolic overload, but rather due to chronic volume deprivation.

Conflict of interest statement

The authors have no conflict of interest to declare.

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