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EDITORIAL

The Fontan Procedure: Now What?

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The decision to commit a patient with single ventricle to the Fontan procedure route is made every day by pediatric cardiologists and cardiovascular surgeons. At this point, there is no other viable alternative to the palliative procedure we currently refer to as Fontan Procedure, first described by François Fontan in 1971 [1], yet more than four decades later there is concern regarding the long-term outcome of patients with Fontan circulation [4, 11, 13].

Specialists in our field will agree regarding the wisdom of choosing the Fontan procedure for patients with single ventricle (anatomical or physiological due to inability to partition), the alternative is a short and complication fraught life of a systemic-pulmonary arterial shunt. Cardiac transplantation is recommended by a limited number of specialists in this field and deemed by most a fate worse than that encountered through lifelong Fontan circulation and reserved by most cardiologists for those with failed Fontan circulation.

The physiological concept behind the Fontan circulation is simple: allow blood to flow from the systemic veins through the pulmonary circulation and beyond to the systemic (single) ventricle without the aid of a pumping (right) ventricle. The flow of blood from the systemic veins to the systemic ventricle is enabled by elevation of the systemic venous pressure, the single ventricle's diastolic relaxation, and the hopefully low resistance of the pulmonary circulation in between. This achieves separation of the two circulations, thus eliminating cyanosis. It also underloads the single ventricle which after completion of the Fontan procedure would have to deal with the systemic cardiac output alone, therefore, reducing the chances of pump failure. Many Fontan patients are alive in their 3rd decade of life, but their wellbeing is challenged by serious complications, many are life-threatening [8].

Fontan procedures completed over the past 3–4 decades were not performed as currently done. It is safe to assume that these surgeries will not be done in the future as currently performed. Surgical techniques coupled with anesthesia and post-operative care allowing safer transition to the new anatomy and physiology of the various phases of the Fontan circulation are constantly improving, therefore, comparing results from yesteryears to current ones is rife with inaccuracies. In addition, interventional cardiac procedures in the cardiac catheterization laboratory currently allow us to improve upon what surgery offers and will certainly add to the better outcome for patients with single ventricle. The fruits of this advancement are noted in many studies of this population of patients assessing ventricular function and cardiac arrhythmias [5, 9], however, the outcome of patients with the Fontan circulation continues to be concerning as unique complications plague these patients, such as heart failure with preserved ventricular contractility, thromboembolism, protein-losing enteropathy, and liver failure. [9, 11, 13]

The shortcomings of a Fontan circulation includes: (a) reliance on a single ventricle and (b) alteration of central venous and pulmonary arterial pressures and flow patterns. The single ventricle has to endure the added burden of increase pulmonary blood flow for months as well as the insult from repeated surgical procedures but thereafter assumes a normal volume burden; this is usually well tolerated if it is architecturally a left ventricle. A morphological right ventricle on the other hand may suffer from inefficient muscular design and an incompetent tricuspid valve. The alteration of the central venous and pulmonary arterial circulations is most probably more

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detrimental than previously thought. The increase in central venous pressure is only one aspect of the changes noted within the Fontan circuit, relative pulmonary hypotension, and lack of pulsatile flow in pulmonary arteries probably play an important role in the disadvantages brought by this type of circulation. It has been postulated that the pathophysiology encountered in Fontan patients may be very similar to that of portal hypertension, where elevated portal venous pressure leads to a cascade of events leading to compensatory increase in hepatic arterial blood flow and steal from the systemic circulation, adding to the "heart failure" manifestations noted in these patients even though the pump mechanism of the single ventricle is preserved [5, 14]. This is supported by Law and colleagues who found that BNP levels were not elevated in Fontan patients with heart failure features but with preserved pump mechanism of the single ventricle (cavo-pulmonary failure [12].

It has been noted [2] that the paradox of increase in central venous pressure and relative hypotension of the pulmonary circulation induced by passive blood flow may lead to elevation of pulmonary vascular resistance. This together with the elevated central venous pressure and loss of normal venous flow pattern in the hepatic veins may be the culprit in Fontan heart failure where the ventricular contractile function is reasonably well preserved. The lack of pulsatility and underfilling of the pulmonary circulation is felt to be responsible for decrease in pulmonary vascular expansion and as such resulting in increased pulmonary vascular resistance which further compounds the problem. It is not clear how the abnormal subdiaphragmatic venous pattern [6] contributes to the pathological changes in liver function and hepatic architecture noted in patients with Fontan circulation, but is likely to be a key pathological aberration which contributes to Fontan circulation failure. These observations by various researchers may point to the significance of the pathological change in venous flow pattern rather than the mere elevation in central venous pressure in patients with these patients.

An important question to consider is: can the Fontan circulation be improved upon to minimize or eliminate the life-threatening complications we have come to know as these patients continue to survive in their 2nd, 3rd decades of life and beyond? If the single ventricular contractile function can be reasonably well preserved through the patients' course of repeated palliative interventions, can the systemic venous/pulmonary circulation pattern be improved upon to provide normal pressures and pulsatility patterns? I am confident that there will be effective measures in these areas; the solution may require better understanding of the cause of liver failure in these patients: is it mere congestions from high central venous pressure, or could it be the lack of variation in venous pressure induced in a normal heart by the right ventricular contractility cycles and process of opening and closure of the tricuspid valve with whatever changes that brings in the venous flow pattern toward the right ventricle. Can the Fontan circulation be saved and allowed to be effective as the patient live beyond their 4th decade? This is possible if the central venous pressure can be kept low and a mechanism duplicating the gentle venous pressure variations through a pulsating (or undulating) mechanism, biological, or mechanical can be devised.

The news from the Fontan population is not all gloom and doom; many studies have shown some reassuring data of these patients as far as rhythm disturbances [10], neurocognitive development [3, 15], and quality of life [7], it is certainly hoped that such good news can spread to all aspects of these patients' health.

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