REVIEW ARTICLE

HEMODYNAMIC PECULIARITIES AND LONG-TERM CONSEQUENCES OF A FONTAN CIRCUIT

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Abstract

Introduction: The Fontan circuit is the corrective circulatory pathway for children with congenital heart defects who have a single functional ventricle. This article reviews the haemodynamic peculiarities of the Fontan circuit and highlights the pathophysiology that contributes to the complications seen in the Fontan physiology.

Keywords: Fontan circuit. Single ventricle, Cavo pulmonary system.

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INTRODUCTION

The Fontan surgical procedure is a rescue intervention done for children born with complex congenital heart malformations who cannot have a biventricular repair of their heart defects. These include tricuspid atresia, hypoplastic left lung syndrome, complicated double outlet right ventricle, heterotaxia (splenic syndromes), complete atrioventricular septal defects, pulmonary atresia with intact ventricular septum and double inlet left ventricle. The complexity of these children's circulatory challenge is that both their systemic and pulmonary circulations are working in parallel and survival only happens if there is an intra-cardiac mixing of both oxygenated and deoxygenated blood.

The Fontan surgical procedure was first performed in 1968 and reported in 1971.[1] Since the introduction of the Fontan to the medical community, a lot of modifications have been made to the procedure but the general principle behind it remains the same and that is, to restore pulmonary blood flow and suppress intracardiac mixing.[1,2] The procedure separates the pulmonary and systemic circulations by bypassing the right heart, which in normal populations provides the driving force to supply the pulmonary circulation. As a result, the pulmonary circulation in the Fontan circuit is driven by small post-capillary pressure. This right heart bypass is achieved by connecting the superior and inferior vena cava directly to the pulmonary artery. The article will discuss the hemodynamic peculiarities and long-term consequences of a Fontan circuit.

THE FONTAN CIRCUIT

Conversion to a full Fontan circulation is done in stages. The first stage is a systemic to pulmonary shunt done soon after birth to provide pulmonary blood flow that is adequate for oxygen delivery. The second stage which is done at 2 to 6 months, is a superior cavopulmonary connection between the superior vena cava (SVC) and the proximal right pulmonary artery. This increases oxygen saturation to 80% from an initial 78%.[3] The third stage is the completion of the Fontan circuit and it involves directing the inferior cava to join the pulmonary circuit via an

intracardiac baffle or an extra cardiac conduit. The outcome is that the whole venous return directly passes to the pulmonary system while all pulmonary vein returns oxygenated blood into the functional ventricle which now supplies the systemic circulation.[4]

The success of the Fontan circulation depends on low pulmonary vascular resistance (PVR) and adequate functioning of the ventricle. The caval venous pressure has to overcome the resisting pressure in the pulmonary vascular bed for blood to flow into the pulmonary vascular bed. Applying Ohm's law to fluid flow means that as the differential pressure between the caval veins and the pulmonary artery increases, blood flow increases while as the PVR increases, blood flow to the pulmonary bed will also decrease. A good ventricular function is also vital to maintain cardiac output, although the determinant of cardiac output in the Fontan circulation is significantly determined by the preload in the functional ventricle. The absence of a pre-pulmonary pump contributing to the propelling energy means that venous return to the pulmonary vascular bed is restricted to the differential pressure in the pulmonary vascular bed and this in turn would control the preloading of the functional ventricle.[5] High pulmonary pressures and atrioventricular valve regurgitation can lead to early failure of the Fontan circuit.[2] Fontan cardiac output has been estimated to be decreased by 70% when compared to that of the normal population.[6] The hallmark of the Fontan circulation is an elevated systemic venous pressure and a decreased cardiac output with a cascade of pathophysiological consequences that affects multiple organs in the body.[7]

THE CAVOPULMONARY SYSTEM

In the Fontan, the pulling of the systemic post-capillary pressures in the SVC and IVC and directly connecting them to the pulmonary capillaries produces a big manmade neoportal system.[2,5] Blood flow through this system supplies the preload to the ventricles and the amount of blood flow that can freely pass through the system is determined by the impedance to blood flow within the system. In the Fontan, this impedance is contributed by surgical cavopulmonary connections, the pulmonary arteries, the pulmonary capillary network, and the pulmonary veins with their atrial connections. This ultimately forms a bottleneck to blood flow in the circuit.[5] The presence of this bottleneck can cause an upstream congestion which is represented by systemic venous congestion in the Fontan circulation and a downstream limitation which is reflected by decreased ventricular preload and as a result decreased cardiac output[5] with diastolic dysfunction that occurs over time.[8] The Fontan circulation can be described as that of chronic systemic venous congestion with resultant higher systemic venous pressure and suboptimal cardiac output.

Systemic veins

The systemic venous pressure has to work to overcome a relatively higher afterload than the right ventricular afterload in normal circulation. A third of the energy generated by the right ventricle is used to maintain patency of the pulmonary artery, the loss of that energy in the Fontan circulation means that the pulmonary arteries are less patent and so contribute more resistance to blood flow which results in a higher afterload for the systemic veins.[9] The high resistance against which systemic veins have to pump produces a state of chronic systemic venous hypertension which can cause peripheral edema, ascites, and lymphatic failure. This pressure can also be transmitted to the portal venous system causing cirrhosis, hepatic dysfunction, and portal venous hypertension.[10]

Exercise tolerance

The ability of patients with Fontan circulation to maximally increase their heart rate during exercise is impaired, thus giving them a low chronotropic index. The reason for this could be due to an intrinsic sinus node dysfunction and the absence of a cardiac pump to increase pulmonary blood flow.[5,11,12] Pulsatile flow is important for the release of endothelium-derived nitric oxide for the recruitment of pulmonary capillary veins and keeping them patent. All of these help to reduce PVR. In the Fontan circuit however, the absence of pre pulmonary cardiac pump means that pulsatile flow is absent and consequently a lack of endothelium-derived nitric oxide and as a result their ability to increase pulmonary blood flow to meet increased metabolic demands during exercise is impaired.[10,12–14] Exercise tolerance in patients with Fontan circuit is 37% lower than that of healthy matched controls.[15]

Arrhythmia

Patients with Fontan circuit have a high risk of developing atrial dysrhythmias. The old Fontan circuit incorporated the right atrium which caused atrial dilatation and hypertrophy, with modern modifications, most patients still have an atriotomy which can cause injury to the sinus node or its conducting fibers and predispose them to arrhythmia.[2,16] Incidence of arrhythmia increases with time in patients with the Fontan circuit and can be up to 50%, this leads to failure of the circuit and is usually associated with high mortality.[17,18]

Lymphatic dysfunction

The Fontan circuit stretches the functional limit of the lymphatic system because the elevated central venous pressure increases tissue fluid and lymphatic fluid production, and at the same time there is a decrease in the drainage of the thoracic duct. This causes a buildup of lymphatic fluid and subsequent leakage of highly proteinaceous lymph into lower-pressure cavities like the intestinal lumen and bronchial airway.[7]

The loss of this protein via the intestinal lumen is termed a protein-losing enteropathy (PLE) and it presents with edema, ascites,

immunodeficiency, malabsorption of fat, hypercoagulopathy, hypocalcemia, and hypomagnesemia. PLE develops in 3.7 to 11% of children by 3 to 7 years after Fontan operation.[19–21] In the bronchial tree, this lymphatic dysfunction manifests as plastic bronchitis where the bronchial tree is filled with casts made up of proteinaceous, and mucoid material with a few lymphocytes. This results in airway obstruction, atelectasis, and expectoration of long, branching bronchial casts. It is potentially very fatal although the incidence is very rare.[22–24]

Conclusion

The Fontan procedure was described by the pioneer surgeon as a physiological blood flow restoration and not an anatomical correction. A low PVR has been identified as the bottleneck of this circuit. With a right ventricular bypass, however, there is exposure of the pulmonary bed to venous pressure, and the absence of a pulsatile pressure from a cardiac chamber would consequently increase pulmonary vascular resistance with time. The Fontan circuit although a good rescue procedure, its efficiency would gradually decline over time resulting in complications. With a growing population of children with operated complex heart diseases having the Fontan physiology, management of these anticipated complications is a growing field.

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