Fontan Repair of Single Ventricle Physiology

Consequences of a Unique Physiology and Possible Treatment Options

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KEYWORDS

• Single ventricle • Fontan • Hepatic dysfunction • Protein losing enteropathy • Heart failure

KEY POINTS

- Understand Fontan physiology.
- Identify Fontan related long-term complications.
- Design an individualized treatment plan.

INTRODUCTION

The Fontan operation is a unique strategy that attempts to create a circulation in series in a patient with single ventricle physiology.[1] Fontan physiology relies on nonpulsatile, passive flow of blood though the pulmonary circulation. Elevation of central venous pressure, elevated pulmonary vascular resistance (PVR), systolic and diastolic dysfunction, and chronic cyanosis can contribute to the phenotype of the "failing Fontan." As the causes of low cardiac output in a patient after the Fontan operation are multifold, treatment plans need to be individualized based on understanding the physiology and the patient's clinical picture. This review provides a summary of Fontan physiology, long-term complications, and potential treatment strategies.

SINGLE VENTRICLE EPIDEMIOLOGY, ANATOMY, AND PHYSIOLOGY

Functional single ventricle anatomy can also result from the inability to septate 2 well formed ventricular chambers.[2,3] These subtypes include, but are not limited to, double inlet atrioventricular (AV) connections, valvular atresia or severe stenosis, and unbalanced AV septal defect. All are associated with a single ventricular cavity, but functional single ventricle anatomy can also result from the inability to septate 2 well-formed ventricular chambers. Given the heterogeneity of the anatomic subtypes, the prevalence of disease can be difficult to determine. Recent data from Marelli and colleagues[4] examining the population in Quebec in 2010 showed a prevalence ratio of univentricular heart to be 0.15 in 1000 in children and 0.05 in 1000 in adults. Hypoplastic left heart syndrome, the most common variant of univentricular heart, had a prevalence ratio of 0.09 in 1000 in children and 0.03 in 1000 in adults.[4]

The clinical presentation and physiology depend on the underlying heart defects; specifically, the presence of outflow tract obstruction, AV valve obstruction or regurgitation, and anomalous venous return determine the type of palliative procedure needed to stabilize the infant. In general, intracardiac mixing across an atrial septal defect and shunting of blood through a patent ductus arteriosus is needed for survival.

Financial Disclosure: The authors have no relevant financial disclosures.
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Cardiol Clin 33 (2015) 559–569
http://dx.doi.org/10.1016/j.ccl.2015.07.010
0733-8651/15/$ – see front matter

Directionality of blood flow across the patent ductus arteriosus is determined by the degree and type of outflow tract obstruction. In the case of hypoplastic left heart syndrome, a patent atrial septum is needed for adequate delivery of oxygenated blood in addition to right-to-left flow across a patent ductus arteriosus to maintain systemic outflow. The goal of initial palliative surgery is to provide stable sources of both pulmonary and systemic blood flow with the eventual goal of separating deoxygenated venous return from oxygenated systemic output.

ANATOMIC AND PHYSIOLOGIC CONSIDERATIONS OF THE FONTAN OPERATION

First described in 1971 by Fontan and Baudet, the Fontan procedure attempts to circumvent the need for a subpulmonary ventricle by redirecting systemic venous return from the superior and inferior vena cava directly to the pulmonary arteries.[1] The classic Fontan operation involved a valved conduit from the right atrium to pulmonary artery, attempting to use the right atrium as a pump to the pulmonary arteries. This evolved to the classic Fontan, which involved a direct anastomosis of the right atrium to the pulmonary artery (**Fig. 1**A). Over time, the right atrium became severely dilated and modifications were proposed to improve hemodynamics and decrease the incidence of arrhythmias. In 1987, de Leval described creating an intraatrial tunnel using the right atrial posterior wall and an end-to-side anastomosis of the superior vena cava to the right pulmonary artery (see **Fig. 1**B).[5] This was followed by the development of an entirely extracardiac conduit directing caval flow to the pulmonary arteries (see **Fig. 1**C).[6] In the newer modifications, a fenestration is often created in the Fontan baffle be creating a small opening between the Fontan pathway and the atrium, allowing for right to left shunting.[7] This shunting allows for augmentation of cardiac output in the early postoperative period, although this is in exchange for the cyanosis that results from the right-to-left shunt. Assessment can be performed as the patient ages to determine candidacy for fenestration closure.[8]

The Fontan operation produces a state of chronic low cardiac output.[9] The Fontan operation relies on nonpulsatile, passive flow of caval blood to the pulmonary arteries. Because there is not a subpulmonary ventricle, the circulation relies on low pulmonary pressures and low PVR. Forward flow through the pulmonary vasculature depends on a differential between the central venous pressure and left atrial pressure. The reasons for progression to the "failing Fontan" physiology are multifold and extend beyond systemic systolic ventricular dysfunction. Caval pressures and the nonpulsatile flow to the pulmonary vascular bed result in chronically elevated central venous pressure, which causes progressive hepatic dysfunction in addition to decreased cardiac output.[10] Increases in pulmonary pressures and PVR only compound these effects as does diastolic dysfunction. Elevation of ventricular end-diastolic pressure can not only cause impaired ventricular filling, but also impairs forward flow through the Fontan baffle and pulmonary arteries. Ventricular systolic dysfunction can occur, but frequently patients with Fontan failure have preserved ventricular systolic function.

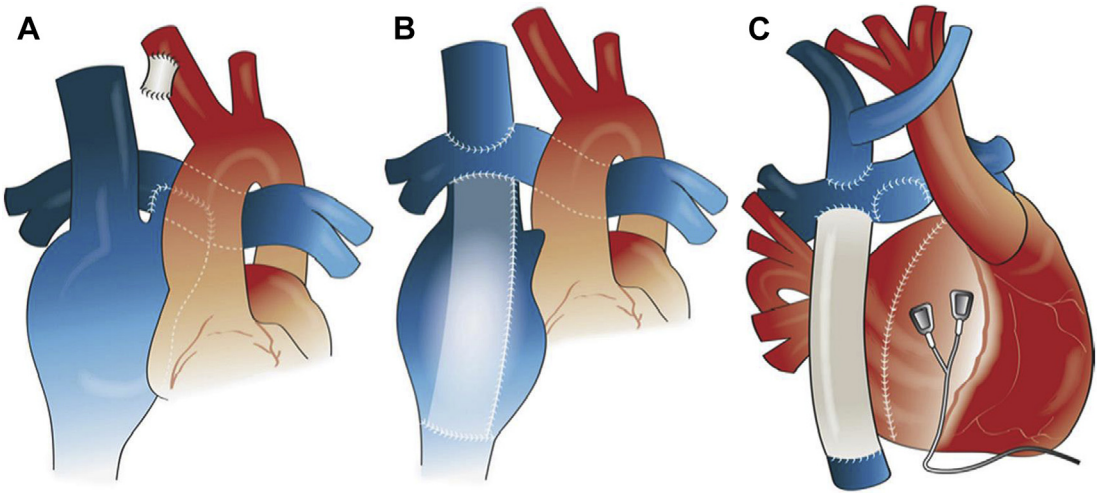

Fig. 1. Variations of the Fontan operation. (*A*) Modified classic right atrium to pulmonary artery Fontan. (*B*) Lateral tunnel Fontan. (*C*) Extracardiac conduit (with extracardiac pacemaker leads). (*From* Khairy P, Poirier N, Mercier L. Univentricular heart. Circulation 2007;115:804; with permission.)