# PERSPECTIVE

Novel Insights into Fontan Procedure: Advancing the Treatment of Congenital Heart Defects

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# Description

Congenital heart defects are among the most common birth defects, affecting millions of people worldwide. Fortunately, medical advancements have revolutionized the management of these conditions, providing improved outcomes and quality of life for patients. One significant breakthrough in the field of pediatric cardiology is the Fontan procedure

Before looking into the Fontan procedure, it is essential to understand the nature of congenital heart defects. These are structural abnormalities present at birth, affecting the heart's structure and function. They can vary significantly in their complexity, ranging from simple defects with minimal impact on health to complex anomalies that can be life-threatening.

In normal hearts, oxygen-depleted blood returns from the body to the right atrium, then flows to the right ventricle and is pumped to the lungs for oxygenation. The oxygenated blood then returns to the left atrium, passes through the left ventricle, and is distributed throughout the body.

In the case of severe congenital heart defects, a complete separation of the systemic and pulmonary circulations is not possible. As a result, oxygen-rich and oxygen-poor blood may mix, leading to reduced oxygen supply to the body and vital organs. The Fontan procedure addresses this problem in a remarkable way.

### Purpose of the fontan procedure

The Fontan procedure is typically performed on children born with single ventricle physiology or other complex congenital heart defects that make it impossible for the heart to function as it normally would. These conditions can include Hypoplastic Left Heart Syndrome (HLHS), Tricuspid Atresia, and Double Inlet Left Ventricle (DILV), among others.

The primary objective of the Fontan procedure is to reroute oxygen-poor blood directly to the pulmonary arteries, bypassing the right ventricle. This separation of blood flow allows for better oxygenation of the blood and improves the overall function of the heart.

The Fontan procedure is typically conducted in a staged manner over several years, as it involves multiple surgeries to gradually reroute blood flow. The overall procedure can be summarized in the following stages.

Stage I-initial surgery: The first stage is often performed soon after birth. It involves redirecting blood from the upper body to the pulmonary arteries through a surgical shunt. This shunt helps improve blood oxygenation by allowing oxygen-poor blood from the upper body to mix with the oxygen-rich blood from the lungs.

Stage II-glenn procedure: This intermediate stage is usually performed when the child is between four and six months old. During the Glenn procedure, the surgeon connects the superior vena cava (the large vein carrying oxygen-poor blood from the upper body) directly to the pulmonary arteries. This bypasses the right ventricle and improves blood flow to the lungs.

Stage III-fontan completion: The final stage, known as the Fontan completion, is typically performed when the child is between two and four years old. In this stage, the surgeon redirects the inferior vena cava (the large vein carrying oxygen-poor blood from the lower body) to the pulmonary arteries. This completes the separation of oxygen-poor and oxygen-rich blood, ensuring that all oxygen-poor blood returning from the body reaches the lungs.

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# Post-procedure management and considerations

After the Fontan procedure, patients require lifelong follow-up care with pediatric cardiologists. While the procedure significantly improves blood flow and oxygenation, it is not a cure for the underlying heart condition. Children who have undergone the Fontan procedure may experience some limitations in physical activity and may require medications to manage potential complications such as arrhythmias or fluid retention.

# **Outcomes and challenges**

The Fontan procedure has undoubtedly been a medical milestone, allowing many children with complex congenital heart defects to survive and lead relatively normal lives. However, it is essential to acknowledge that the procedure is not without challenges. Some children may still experience long-term issues, including heart failure, protein-losing enteropathy, and plastic bronchitis. Ongoing research aims to improve patient outcomes and address these challenges.

The Fontan procedure stands as a testament to the remarkable advancements in pediatric cardiology. It has offered hope and extended lives for countless children born with complex congenital heart defects. As medical knowledge continues to evolve, people can expect further refinements and innovations that will continue to improve the outcomes and quality of life for these young patients, granting them the opportunity to embrace a brighter future.