Failure of the Fontan Circulation

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KEYWORDS

• Fontan circulation • Chronic low output • Circulatory failure • Pulmonary vascular resistance

KEY POINTS

- The essence of a Fontan circuit is the creation of the Fontan "neoportal system": this allows for oxygenation at near normal levels, but at the cost of a chronic state of systemic venous congestion and decreased cardiac output.
- The heart, while still the engine of the circuit, cannot compensate for this major flow restriction: the ventricle has lost control of the output and of systemic venous congestion; systolic and diastolic ventricular dysfunction are common and may contribute to overall circulatory failure.
- The abnormal hemodynamics inherent in the Fontan circulation affect organs outside the heart and may lead to liver cirrhosis, protein-losing enteropathy, or plastic bronchitis.
- Failure of the Fontan is progressive; over time there is an insidious increase in both pulmonary vascular resistance and ventricular end-diastolic pressure, which may lead to progressive functional impairment.

THE "FONTAN" CONCEPT

A normal mammalian cardiovascular system consists of a double circuit, pulmonary and systemic, connected in series and powered by a double pump. In the absence of congenital heart disease, the right ventricle pumps to the pulmonary circulation and the left ventricle pumps to the systemic circulation (**Fig. 1**A).

Many complex cardiac malformations are characterized by the existence of only one functional ventricle. This single ventricle has to maintain both the systemic and the pulmonary circulations, which at birth are not connected in series but in parallel (see **Fig. 1**B). Such a circuit has 2 major disadvantages: diminished oxygen saturation of the systemic arterial blood and a chronic volume load to the single ventricle. The chronic ventricular volume load will lead to progressive impairment of ventricular function and altered pulmonary vasculature, causing a gradual attrition resulting from congestive heart failure and pulmonary hypertension from the third decade, with few survivors beyond the fourth decade.

In 1971, Francis Fontan[1] from Bordeaux, France, reported a new approach to the operative treatment of these malformations, separating the systemic and pulmonary circulations. In a "Fontan circulation" the systemic venous return is connected to the pulmonary arteries without the interposition of a pumping chamber (see **Fig. 1**C). In this construct, residual postcapillary energy is used to push blood through the lungs in a new portal-like system.[2] Advantages of a Fontan circuit include (near) normalization of the arterial oxygen saturation, and abolishment of the chronic volume load on the single ventricle. However, because venous return through the pulmonary vasculature

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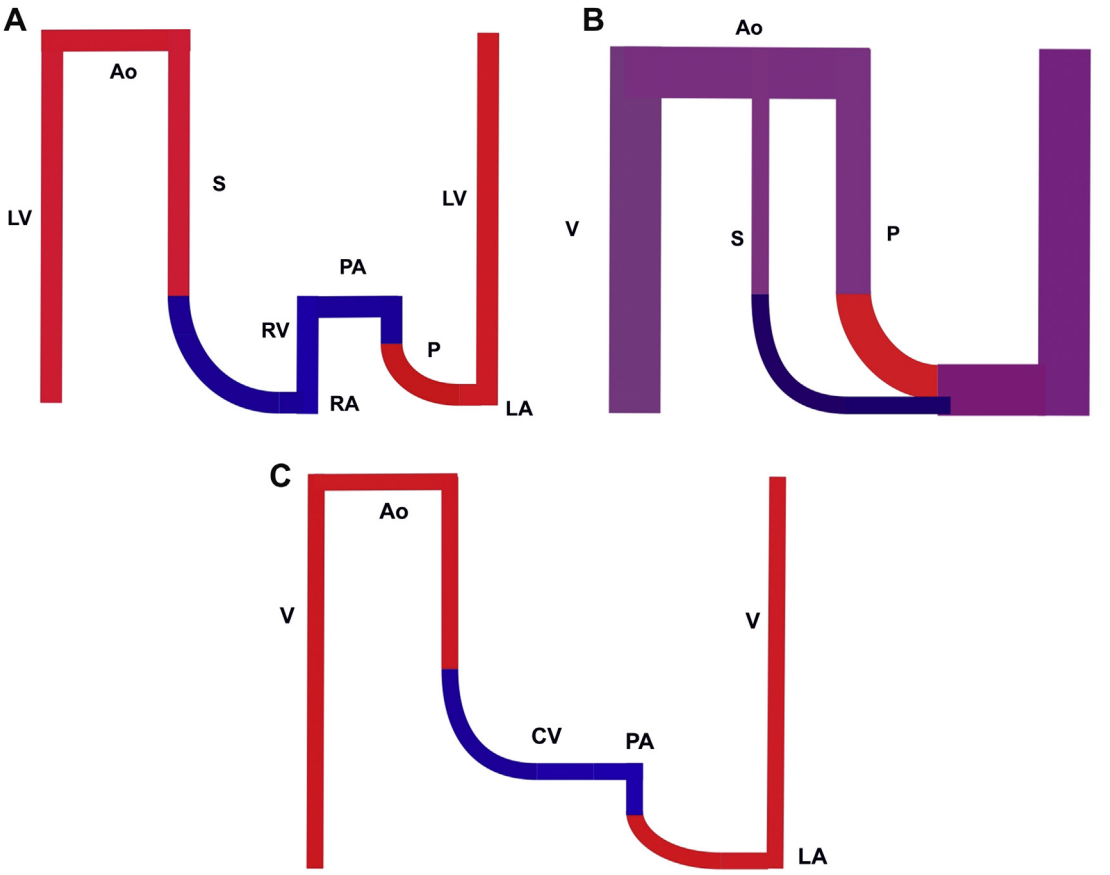

Fig. 1. Normal cardiovascular circulation (*A*), shunted palliation (*B*), and Fontan circulation (*C*). (*A*) Normal circulation. The pulmonary circulation (P) is connected in series with the systemic circulation (S). The right ventricle maintains the right atrial pressure lower than the left atrial pressure, and provides enough energy for the blood to pass through the pulmonary resistance. (*B*) Shunted palliation. The systemic (S) and pulmonary (P) circuits are connected in parallel, with a considerable volume overload to the single ventricle. There is complete admixture of systemic and pulmonary venous blood, causing arterial oxygen desaturation. (*C*) Fontan circuit. The systemic veins (V) are connected to the pulmonary artery, without a subpulmonary ventricle or systemic atrium. The lungs are thereby converted into a neoportal system, which limits flow to the ventricle. In the absence of a fenestration, there is no admixture of systemic and pulmonary venous blood, but the systemic venous pressures are markedly elevated. A fenestration allows the systemic venous blood to bypass the Fontan portal system and limits the damming effect, thereby increasing output and decreasing congestion, but also arterial saturation. Ao, aorta; CV, caval veins; F, fenestration; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle; V, single ventricle. Line thickness reflects output, color reflects oxygen saturation.

is hindered by the pulmonary impedance, this circulation creates a state of chronic hypertension and congestion in the systemic veins, and results in decreased cardiac output, both at rest and during exercise (**Fig. 2**).[3,4] It is these 2 inherent features of the Fontan circulation, elevated systemic venous pressure and chronically low cardiac output, which are the root cause of most of the physiologic impairments, collectively termed Fontan failure.

CARDIAC OUTPUT IN THE FONTAN CIRCULATION

By creating a total cavopulmonary connection, a new portal system is made. A portal system occurs when one capillary bed pools blood into another capillary bed through veins without passing through the heart; for example, the hepatic portal system and the hypophyseal portal system. The Fontan neoportal system dams off and pools the systemic venous blood. As a result, transit of