The Failing Fontan: Etiology, Diagnosis and Management
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The Fontan operation results in separation of the circulations with direction of the deoxygenated systemic venous return to the lungs absent a pulmonary ventricle. It is the anticipated ultimate stage of surgical reconstruction for single ventricle, which leads to an increase in oxygen saturation. However, the physiology created is suboptimal when compared to normal, with a number of serious physiological limitations. Passive venous return to the lungs without a pumping chamber leads to elevated central venous pressure. There is an impaired ability to deliver a normal quantity of blood volume across the pulmonary vascular bed. Abnormalities of the pulmonary vasculature can contribute to impediment to passive forward flow. This results in decreased ventricular filling, with low stroke volume and hence low cardiac output at rest. There is also the inability to adequately increase stroke volume and cardiac output during periods of exercise. Chronotropy is impaired as a consequence of the surgery itself, which further leads to diminished cardiac output. Systemic vascular resistance is increased. Ventricular dysfunction, both systolic and diastolic is common, as the architecture and performance of the single ventricle varies from the normal systemic left ventricle.

The patient after Fontan operation essentially exists in a chronic low cardiac output state with elevated central venous pressure. These physiological limitations are manifested through a number of clinical conditions. In this lecture, we will discuss our current understanding of protein losing enteropathy and plastic bronchitis, and review treatment strategies for these ailments.