Many congenital heart defects result in univentricular physiology, having in common anatomical features that preclude surgical septation of the ventricles. Although these defects have variable clinical manifestations in childhood, the common endpoint of staged surgical palliation is the Fontan procedure. The concept that the right ventricle could be bypassed allowing systemic blood to enter the pulmonary artery directly was first established on experimental animals in 1970. A right atrial to pulmonary artery surgical procedure in humans with tricuspid atresia was introduced almost simultaneously by Fontan and Kreutzer in 1971. Over the years, the indications for the Fontan operation gradually expanded to all patients with single ventricle physiology.

Surgical techniques used to perform the Fontan procedure have evolved several times since it was first described. Regardless of the exact nature of the connection, the completed circulation is often described as one having a single energy source, the systemic ventricle. This energy is dissipated through a series of resistors: in the ventricle itself (related to diastolic function), the systemic vascular bed, the systemic venous bed and the pulmonary vascular bed. Despite the new era of survival and potential that the Fontan circulation offers such patients; this unique circulation is abnormal in virtually every aspect of its performance.