

More people born with a single lower heart chamber survive; but, face challenges in quality and length of life

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The Fontan procedure has allowed more people born with only one ventricle—the lower pumping chamber of the heart—to survive into adulthood, but their unique circulatory system requires continued

lifelong medical care. A new scientific statement from the American Heart Association, published in the American Heart Association journal *Circulation*, summarizes the current state of knowledge on Fontan circulation and how best to care for these unique patients.

There are two ventricles in a normal heart—one pumps blood to the lungs and the other pumps blood to the rest of the body. In children born with only one [ventricle](#), a surgical procedure (the Fontan procedure) diverts blood returning from the veins directly to the main pulmonary artery leading to the lungs, instead of being pumped from the heart.

Typically, people with Fontan [circulation](#) have chronically elevated pressure in their veins and less blood being pumped out of their heart. This can lead to circulatory failure because when the heart pumps less efficiently it is not able to provide enough oxygen to the cells in the body. In addition, these [patients](#) often experience ventricular dysfunction, heart failure, heart rhythm disturbances and problems with their liver, kidneys, bones and other organ systems.

The statement provides recommendations for follow-up care for patients with Fontan circulation, including guidance on strategies for maintaining the health of the [heart](#) and organs through "surveillance testing—routine, systematic evaluation of both cardiovascular and other organs affected by Fontan circulation.

Gaps in knowledge and areas for future investigation are also highlighted, with the objective of laying the groundwork for creating a normal quality and duration of life for these unique individuals. "We need more research into the basic biology of single ventricle hearts and whether the damage to other [organ systems](#), such as kidneys, liver and brain can be mitigated or reversed," said writing group chair Jack Rychik, M.D., Robert and Dolores Harrington endowed chair in cardiology and Professor of Pediatrics, at the Children's Hospital of

Philadelphia.

Although life expectancy for people born with one ventricle is lower than average, people with Fontan circulation can live a rich and fulfilling life.

"We are entering a new phase in the management of patients born with one ventricle. Provided that patients undergo regular follow-up with their healthcare provider, adopt a [healthy lifestyle](#) and are encouraged to participate in investigational clinical protocols and research, [healthcare providers](#) and patients can share an optimistic and hopeful view for a brighter future," Rychik said.

The worldwide population of patients with Fontan circulation grew to an estimated 50,000 to 70,000 patients in 2018, with 40% of patients aged 18 years or older.

"Patients with Fontan circulation are going to consume an ever-increasing amount of resources as they grow in number and age into adult life. Healthcare providers, both pediatric and adult, will need to increase their understanding and knowledge of this unique cardiovascular condition in order to maintain and improve their quality of life" Rychik said.

More information: Jack Rychik et al. Evaluation and Management of the Child and Adult With Fontan Circulation: A Scientific Statement From the American Heart Association, *Circulation* (2019). DOI: 10.1161/CIR.0000000000000696 , www.ahajournals.org/doi/10.1161/CIR.0000000000000696

Provided by American Heart Association

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