

UNIVENTRICULAR HEART

(Tricuspid atresia/Single ventricle)

Definition

Univentricular heart denotes malformations where either the RV or LV is missing or, if present, is hypoplastic, and thus not amenable for biventricular repair, such as:

- Tricuspid atresia
- Mitral atresia
- Double-inlet left or right ventricle
- Hypoplastic left or right heart syndrome variants
- Heterotaxia syndromes

These malformations are always associated with additional intracardiac and extracardiac lesions.

Pathophysiology and presentation

The following haemodynamic conditions can be seen:

1. Patients with no anatomic restrictions to pulmonary blood flow with early postnatal development of a large left-to-right shunt and symptoms of congestive heart failure. This condition may be exacerbated by obstructions to systemic blood due to additional abnormalities. Surgical treatment, such as coarctation repair and pulmonary artery banding is usually needed early in life to avoid development of severe pulmonary vascular disease.
2. Patients with severe cyanosis due to obstruction to pulmonary flow, frequently caused by valvular or subvalvular PS or atresia. A small number of patients, usually with the right-isomerism type of heterotaxia, can also have total anomalous pulmonary venous connection. These patients usually undergo a systemic to pulmonary artery shunt procedure, with repair of the total anomalous pulmonary venous connection if needed.

A small number of patients will present with mild cyanosis and no congestive heart failure without a previous operation. Usually, adults with these conditions will have undergone previous palliation with some type of a systemic to pulmonary shunt.

Therapy

Recommendations for management of these patients are presented in Table 1. A discussion of management following Fontan repairs is provided in the relevant Chapter. Depending with haemodynamic status, single ventricle anomalies are initially dealt with PA banding or a Norwood procedure (modified Blalock-Taussig or RV to PA shunt) and later on with a

Fontan operation.¹ In the SVR trial (Single Ventricle Reconstruction), 1-year transplant-free survival was better for the Norwood procedure with right ventricle-to-pulmonary artery shunt compared with a modified Blalock–Taussig shunt in patients with hypoplastic left heart and related syndromes, but at 6 years the hazards of death or transplant and catheter interventions were not different.² With contemporary surgical and medical advances, >85% of individuals who undergo Fontan palliation now survive into adulthood, and >70 000 people worldwide are estimated to be living with Fontan circulation.³ Among survivors of the Fontan procedure, complications and the need for additional interventions increase over time, but cardiac transplant-free survival, up to 90% over 12 years, is independent of ventricular morphology or age.⁴ The risk for adverse cardiovascular events, including all-cause mortality, cardiac transplantation, and atrial arrhythmias is more than 2-fold higher in patients with univentricular hearts with a dominant morphological right vs left ventricle following Fontan palliation.⁵ Despite improved surgical techniques, preoperative and postoperative care, patients with functionally univentricular hearts continue to have considerable morbidity and mortality and a decreased lifespan.⁶ Mental health and neuropsychologic care are essential components of routine multidisciplinary Fontan care.³

References

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2. Newburger JW, et al. Transplant-Free Survival and Interventions at 6 Years in the SVR Trial. *Circulation*. 2018;137:2246-2253.
3. Glenn TM, et al. The Fontan Outcomes Network: Findings After 2 Years and 1121 Participants. *Circulation*. 2026;153:1182-1195.
4. Atz AM and al. e. Longitudinal Outcomes of Patients With Single Ventricle After the Fontan Procedure. *Journal of the American College of Cardiology*. 2017;96.
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