

Long-term Mortality risk after Fontan Procedures: A systematic review.

Abstract:

Background

Despite an aging Fontan population, data on late outcomes is still scarce. Reported outcome measures and determinants vary greatly between studies making comprehensive appraisal of mortality hazard challenging.

Methods

We conducted a systematic review to evaluate causes and risk factors for late mortality in patients with Fontan circulation. Late mortality was defined as mortality beyond the first postoperative year. Studies were included if they had ≥ 90 patients or ≥ 20 late mortalities and/or transplants. The pooled hazard ratios were used to create a clinical score for risk stratification in Fontan patients.

Results

From 28 studies, a total of 9502 patients with an average follow-up time of 8.28 ± 5.07 years were identified. There were 1408 deaths and 138 heart transplantations. Causes of late death were reported in 969 cases. The five most common causes were heart/Fontan failure (25%), arrhythmia, respiratory failure, renal disease and thrombosis/bleeding. Risk factors for late mortality were evaluated and classified into 9 risk categories. The Fontan risk score (ranging from 0-100) includes 9 risk factors including anatomic risk factors, elevated preoperative pulmonary artery pressure, age > 7 at the time of the operation, atriopulmonary Fontan, heart failure symptoms, arrhythmia, moderate/ severe ventricular dysfunction or atrioventricular valve regurgitation, protein losing enteropathy and end organ disease (cirrhosis or renal insufficiency).

Conclusion

Factors associated with late mortality after the Fontan operation are summarized in this study.

The proposed risk score may aid risk stratification in outpatient setting and thus enable the clinician to tailor surveillance to patients' risk profile.

Table 1. Fontan Risk Score.

Risk factor	Number of papers	Hazard ratio range	Final score
Anatomic risk factors (hypoplastic left heart or heterotaxy)	4	2.8-12.7	12
Elevated preoperative pulmonary artery pressure >15 or Fontan pressure >20 mmHg postoperatively	5	1.14-3.5	5
Age > 7 at the time of the operation	2	2.7-2.9	9
Atriopulmonary Fontan	3	3.7-6.2	17
Heart failure symptoms or need for diuretic use	3	1.58-9.16	6
Arrhythmia	2	1.79-6	8
Moderate/ severe ventricular dysfunction or moderate/ severe AV valve regurgitation	2	4.02 -7.17	23
Protein losing enteropathy	5	1.97-8.5	8
End organ disease including cirrhosis or renal insufficiency	4	2.49-19	12

Table 2. Summary of risk factors classified into categories.

Risk factor category	Identified risk factors
Era effect	Fontan operations performed prior to 1990
Pre-operative	HLHS, age at Fontan surgery > 7yrs, PAP >15 mm Hg, poor ventricular function, presence of AV valve regurgitation or heterotaxy syndromes.
Operative	Atriopulmonary Fontan and AV valve replacement at the time of Fontan procedure
Post-operative	Prolonged chest tube drainage, elevated Fontan pressure > 20 mm Hg and left atrial pressures > 13 mm Hg.
Cardiovascular complications	Arrhythmia, heart/Fontan failure and thromboembolic events
Non-invasive cardiac imaging	Lower global longitudinal strain and elevated ventricular end diastolic volume index > 125 cc/m ² on cardiac MRI
Exercise stress test	Poor peak heart rate < 122 BPM and low VO ₂ max < 16.6 ml/kg/min
Non-Cardiac complications	Protein losing enteropathy, plastic bronchitis, Fontan associated cirrhosis and high altitude.
Serologic factors	Hyponatremia < 136 mmol /L and elevated BNP

HLHS indicates Hypoplastic Left Heart Syndrome; PAP, Pulmonary artery pressure; AV, Atrioventricular; BPM, Bear per minute; VO₂ Max, Maximal oxygen consumption, BNP, Brain natriuretic peptide.