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**Table E1: List of available treatment modalities for the “failing” and “failed” Fontan circulation**

|  |
| --- |
| **Early Fontan failure**   * Rapid recognition and correction of any residual surgically correctable anatomic problems. * Terminate tachycardia * Other options: Fontan takedown to bi-directional Glenn / Fontan pathway fenestration / hemi-Fontan / extracorporeal membrane oxygenation or urgent cardiac transplantation * Ventricular assist device including Berlin heart- unsuitable optionE88-E90,E118 |
| **Systemic ventricular dysfunction**   * Often refractory to medical therapy * Afterload reduction, beta blockers inotropesE52 * Acetylcholine esterase- inhibitory drugs46,E52 * Pulmonary vasodilatorsE175,E183-E185 * Calcium channel blockers for diastolic dysfunction * Multisite pacingE37-E39,E119 * Cardiac transplantationE88-E103,E107,E108 |
| **Ascites**   * In early Fontan failure- urgent correction of residual anatomic problems, if any * High risk Fontan- fenestration of Fontan pathway / Fontan takedown to an intermediate circulationE176 * Selected cases- Fontan conversion with arrhythmia surgery, dual chamber pacing20,21,E179 |
| **Lymphatic dysfunction**  *Protein losing enteropathy*   * Multi-nutritional support with protein, median chain triglyceride * Control supraventricular arrhythmias * Afterload reduction, diuretics, * Enteric steroids- oral budesonideE86,E170 * Unfractionated heparinE170,E171 * High dose aldactoneE171,E172,E173 * Intravenous albumin and gammaglobulin infusion * Immunosuppression and targeted pulmonary vasodilators- sildenafilE120,E125 * LoperamideE174 * Exclude Fontan pathway obstruction, * Other options- Atrial pacingE54, Fontan pathway fenestrationE176,E324 * Surgical ligation of the thoracic duct2,3,7,E175,E177 * Interruption of lympho-intestinal connectionsE321 * Cardiac transplantationE100,E107,E176,E184   *Plastic bronchitis*   * Urgent bronchial lavage using inhaled tissue plasminogen and vest therapy to mobilize bronchial castsE181,E182 * Pulmonary vasodilators, aldosterone inhibition, inhaled ileoprost E10,E11,E272-E274 * Percutaneous lymphatic interventions, embolization of lymphobronchial connectionE74,E238 * Surgical thoracic duct ligationE322 * Cardiac transplantationE184 |
| **Lungs**  *Pulmonary remodeling and rising pulmonary vascular resistance*   * Pulmonary vasodilatorsE175,E183-E185 * Inhaled ileoprostE10,E11 * Stenting of pulmonary arterial narrowing, if any |

**Table E2: List of available treatment modalities for the “failing” and “failed” Fontan circulation**

|  |
| --- |
| **Pulmonary arteriovenous malformations**   * Options- percutaneous closure using gel foam coils * Surgical revision of the Fontan circuit to promote uniform blood flow to both lungs ensuring supply of hepatic factor * Cardiac transplantationE88-E103 * Lung transplantation for diffuse and extensive arteriovenous malformationsE180 * Creation of a brachial or axillary arteriovenous fistula to deliver the hepatic factor, and resolve pulmonary arteriovenous malformationsE323 |
| **Cyanosis and collateral formation**   * Options- coil embolization of collaterals * Catheter-based occlusion of the fenestration or residual atrial septal defect9,17,46,E135-E137 * Classic Glenn with discontinuous pulmonary artery- restore continuity * Fontan conversion- selected cases of atriopulmonary connection9,17,20,21 |
| **Hepatic effects**   * Supportive care, optimize cardiac output * Pulmonary vasodilators and angiotensin-converting enzyme inhibitors - therapeutically potential benefit * Cardiac transplantation to reverse cirrhosis of liver, subclinical hepatic disease challenging post cardiac transplantationE49 * Failed Fontan with advanced cirrhosis of liver- cardiac and hepatic transplantationE149,E150,E178 |
| **Hepatorenal insufficiency**   * Supportive care * Optimise cardiac output * High mortality |
| **Fontan pathway obstruction, valve dysfunction, and outflow obstruction**   * Early repair of atrioventricular valves, left ventricular outflow tract obstruction on an individualized basis (Mavroudis, Honja, Kotani)5,6,E22,E37-E39 * Focal pulmonary artery stenosis/pulmonary venous obstruction- revision of Fontan pathway2-7,E186-E188 |
| **Thrombotic pathway obstruction**   * Early Fontan failure- Fontan takedown to an intermediate circulation, allograft replacement of systemic conduit using an allograftE186-E188 * Post-cardiac transplantation- problems of silently elevated pulmonary vascular resistance secondary to silent pulmonary microemboliE100 |

**Table E3: Flow diagram depicting the management algorithm for management of post Fontan supraventricular arrhythmias**

**(Deal BJ, 2007)**

**Acute/immediate evaluation**

* Achieve stable hemodynamics
* Control heart rate, antiarrhythmics (Beta blockers, Amiodarone)
* Terminate tachycardia
* Administer anticoagulation

Assess Fontan circulation

* Advanced imaging
* Hemodynamic evaluation
* Measure metabolic / electrolyte parameters

Intervene prior to ascites / protein losing enteropathy / thrombosis

|  |  |  |
| --- | --- | --- |
| Atriopulmonary or Atrioventricular  **↓**  Fontan conversion and Arrhythmia surgery | Total cavopulmonary connection | |
|  | Unstable hemodynamics  **↓**  Catheter or surgical interventions | Stable hemodynamics, unsuitable or high risk for arrhythmia surgery  **↓**  Catheter ablation |
|  |  |  |

**Surgical options**

* Fontan revision, pulmonary arteroplasty8,17-22,E137-E139,E179
* Fontan conversion to extracardiac Fontan circuit, appropriate modified Maze procedure, routine dual chamber pacing20,21
* Multisite pacing for dyskinetic systemic ventricles20,21

**Definitions (Electronics)**

**Fontan Failure**

Fontan failure **has** been variably and inconsistently described in the literature leading to challenges in comparing studies and evaluating treatments.8-12 Fontan failure is a clinical syndrome where circulation can no longer meet the metabolic demands of body, with symptoms of exercise intolerance with or without congestion resembling heart failure.8,9

The term ‘Failing Fontan’ refers to a clinical situation with major rhythm disturbances refractory to maximal medical therapy, thrombotic events in Fontan circuit, protein losing enteropathy, plastic bronchitis, chronic edema, ascites, cirrhosis, and ventricular failure.8-14 Intervention in failing Fontan may optimize clinical status while maintaining the Fontan circulation.8-16

The “failed Fontan” circulation includes hemodynamic or multiorgan system complications of Fontan circulation which are not reversible by surgical or catheter interventions at acceptable risk. Options for “failed Fontan” are limited to cardiac transplantation or attempts to minimize the impact of irreversible functional deterioration.9,17-21

Early Fontan failure is a postoperative state of low systemic perfusion characterized by high Fontan circuit pressures and large volume requirements, resistant to medical management necessitating the use of extra-corporeal membrane oxygenation, Fontan revision or takedown to an intermediate circulation, emergent cardiac transplantation within 30 days of Fontan procedure or death.15,16

Late Fontan failure may present insidiously over years. It is a failure of medical management to interpret the absence of overt symptoms or ascites as evidence of optimal hemodynamic status in the functionally univentricular circulation.2,9,19 The term ‘Fontan revision’ includes patients undergoing Fontan reoperations and address hemodynamic problems without arrhythmia surgery.17,20,21 The term ‘Fontan conversion’ accounts for the three major parts of new Fontan reoperation, namely, revision of atriopulmonary to extracardiac Fontan connections, atrial arrhythmia surgery, and pacemaker implantation.17,20,21