

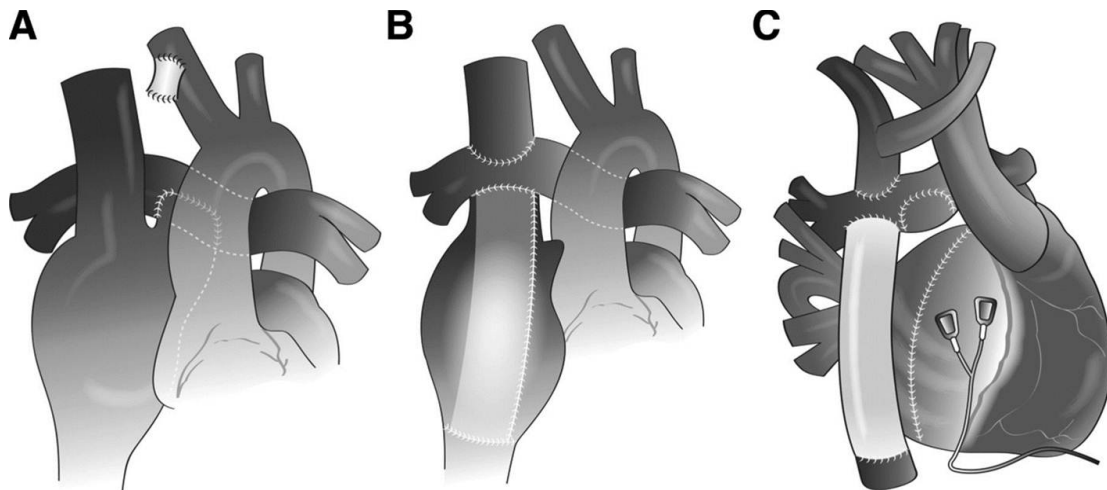


Figure 1. Variations of Fontan surgery.

A, The modified classic Fontan. B, The intracardiac lateral tunnel Fontan. C, The extracardiac.

Khairy P, Poirier N. Is the extracardiac conduit the preferred Fontan approach for patients with univentricular hearts? the extracardiac conduit is not the preferred Fontan approach for patients with univentricular hearts. *Circulation*. 2012;126:2516-25.

Table 1. ACC/AHA 2025 GL on ACHD. Recommendations for Fontan Palliation of Single Ventricle Physiology

Diagnostic	
In adults with Fontan circulation and evidence of progressive Fontan circulatory failure, formal evaluation by a heart failure/transplant cardiologist with experience in heart transplantation in the adult Fontan population is recommended.	1 B-NR
In adults with Fontan circulation, imaging and laboratory evaluation of the liver should be performed at least annually to screen for hepatocellular carcinoma and for evidence of progressive Fontan-associated liver disease (FALD).	1 C-LD
In adults being considered for Fontan palliation or Fontan conversion, cardiac catheterization should be performed to ensure the presence of acceptable hemodynamics.	1 C-EO
In adults with Fontan circulation and new-onset or worsening atrial tachyarrhythmias, evaluation with imaging and follow-up cardiac catheterization as indicated is recommended to assess for potential contributory anatomic or hemodynamic abnormalities and Fontan pathway thrombosis.	1 C-EO
In adults with Fontan circulation and new or progressive symptoms, hypoxemia, declining functional status, or evidence of progressive or newonset noncardiac organ dysfunction, hemodynamic evaluation with cardiac catheterization is recommended to guide therapy.	1 C-EO
In adults with Fontan circulation and newly progressive or severe hypoxemia or hypotension, evaluation with advanced cardiac imaging (MR, CT, transesophageal echocardiography) is recommended to rule out thrombus or emboli in the Fontan or pulmonary vasculature.	1 C-EO
In adults with Fontan circulation undergoing CT angiography to rule out thrombus or emboli in the Fontan or pulmonary vasculature, imaging should be performed using protocols to avoid false-negative or false-positive results.	1 C-EO
In adults with Fontan circulation, annual laboratory evaluation is recommended to assess for evidence of organ-system dysfunction or hematologic abnormality.	1 C-EO
In adults with Fontan circulation, consultation by a hepatologist, in collaboration with an ACHD cardiologist, is reasonable to facilitate interpretation of hepatic testing, diagnose and treat complications related to portal hypertension, and participate in timing and management decisions specific to organ transplantation.	2a C-EO
In adults with Fontan circulation, liver biopsy can be helpful to delineate the degree of hepatic fibrosis or cirrhosis before consideration of transplantation.	2a C-LD
In adults with Fontan circulation undergoing cardiac catheterization	2b C-LD

(diagnostic, interventional, or electrophysiological), it may be reasonable to exclude Fontan-related thrombus before catheter placement to avoid pulmonary or systemic thromboembolism.

Routine Follow-Up and Testing Intervals

Type of Follow-Up or Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	N/A	12	6-12	3-6
Electrocardiogram	N/A	12	6-12	6-12
Transthoracic echocardiogram	N/A	12	12	6-12

Table 2. ACC/AHA 2025 GL on ACHD. Recommendations for Fontan Palliation of Single Ventricle Physiology

Therapeutic

In adults with Fontan circulation and a known or suspected thrombus, atriopulmonary Fontan, a history of thromboembolism, or a history of sustained atrial flutter or fibrillation, anticoagulation is recommended to reduce the likelihood of thromboembolic disease.	1 B-NR
In adults with Fontan circulation and the absence of high-risk features (history of thromboembolism, sustained atrial flutter/fibrillation, or atriopulmonary Fontan) or bleeding contraindications, treatment with either aspirin or anticoagulation is recommended to reduce the probability of thromboembolic disease.	1 B-NR
In adults with Fontan circulation and sinus node dysfunction requiring pacemaker placement, atrial-based pacing with programming to minimize ventricular pacing is recommended to improve patient symptoms and to avoid pacing-induced cardiomyopathy.	1 B-NR
In adults with Fontan circulation with new-onset atrial flutter or atrial fibrillation, timely cardioversion (pharmacological or electrical) is recommended to prevent clinical decompensation.	1 C-LD
Adults with Fontan circulation referred for cardiac transplantation should undergo pretransplantation review by a committee with broad multidisciplinary representation to improve patient selection and posttransplant outcomes.	1 C-EO
In adults with Fontan circulation and atrioventricular block whose burden of ventricular pacing is likely to be high (ie, >40%), apical site pacing is preferred over nonapical sites to improve transplant-free survival.	2a B-NR

In adults with Fontan circulation, formal exercise programs and/or cardiac rehabilitation (appropriate to the patient's ability) can be beneficial in improving functional capacity.	2a B-NR
In adults with Fontan circulation and recurrent supraventricular tachycardia, catheter ablation performed by an electrophysiologist with experience in ACHD is reasonable to reduce arrhythmia recurrence and associated morbidity.	2a C-LD
In adults with Fontan circulation and hemodynamically significant stenosis of the Fontan or PAs, catheter-based stenting of the stenosis to reduce Fontan pressure and improve flow is reasonable to improve hemodynamics.	2a C-EO
In symptomatic adults with Fontan circulation, or those with evidence of systemic ventricular volume overload, closure of aortopulmonary collaterals to improve symptoms and reduce the likelihood of pathologic ventricular remodeling can be useful.	2a C-EO
In adults with Fontan circulation, treatment with pulmonary vasodilators may be considered to improve exercise capacity.	2b B-R
In adults with an atriopulmonary Fontan, preserved ventricular function, and acceptable surgical risk with atrial arrhythmias refractory to medical and catheterbased therapies, Fontan conversion surgery may be considered to reduce arrhythmia recurrence.	2b C-LD
In adults with Fontan circulation and symptoms or Fontan circulatory failure attributable to significant intracardiac structural or anatomic abnormalities, reoperation may be considered to improve symptoms or reduce the likelihood of progressive Fontan circulatory failure.	2b C-EO

Table 3. ESC 2020 GL on ACHD. Special considerations and recommendations for intervention after Fontan operation

Electrical cardioversion for sustained atrial arrhythmia with rapid AV conduction (medical emergency)	I C
Anticoagulation in the presence, or with a history, of atrial thrombus, atrial arrhythmias, or thromboembolic events.	I C
Women with a Fontan circulation and any complication are counselled against pregnancy.	I C
Cardiac catheterization at a low threshold in cases of unexplained oedema, exercise deterioration, new-onset arrhythmia, cyanosis, and haemoptysis.	I C
A proactive approach of electrophysiologic evaluation and ablation (where appropriate) in patients with arrhythmias.	IIa C
Regular liver imaging (ultrasound, computed tomography, magnetic resonance)	IIa C
Endothelin receptor antagonists and phosphodiesterase- 5 inhibitors in selected patients with elevated pulmonary pressure/resistance in the absence of elevated ventricular end diastolic pressure.	IIb C
In selected patients with significant cyanosis, device closure of a fenestration, but requires careful evaluation before intervention to exclude induction of systemic venous pressure increase or fall in cardiac output.	IIb C

AV = atrioventricular.

Table 4. 2025 ESC 2025 Guidelines on pregnancy.

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
Single ventricle physiology palliated with Fontan circulation				
	<i>High pregnancy risk</i> CV events⁴⁷⁹: - Supraventricular arrhythmias (8%–11%) - HF (4%–14%) <i>Risk factors:</i>⁴⁷⁹ - Oxygen saturations <85% ↓ Ventricular function - Arrhythmias - Significant valvular disease - NYHA class III/IV - FALD	↑ <i>Very high foetal complication risk:</i>^{479–483} - Low live birth rate (56%) - Miscarriages (45%–54%) - SGA (20%–55%) - Premature birth (59%–72%) - Neonatal death (5%–18%) <i>Obstetric risk:</i> - Hypertension (14%) - PPH (13%)	- According to anatomical and functional status: TTE every 1–3 months and serial NP⁴⁸⁴ - FU in specialized ACHD centre	- Pre-pregnancy counselling about very high risk especially if any risk factor (see maternal risk) - Pregnancy may be well tolerated and successful in a subset of women with single ventricle and Fontan circulation without complications^{479,480,485–487} - ASA and/or LMWH (depending on the presence of complications) should be considered in shared decision⁴⁸⁴ - Atrial tachyarrhythmias should be promptly treated with cardioversion¹ <i>Labour/delivery with preload dependent circulation:</i>⁴⁶¹ - Epidural with slow titration - Labour in left lateral decubitus position - Low thresholds for assisted second stage (↓ Valsalva duration) iv. air filter (if fenestration or significant venovenous collaterals)