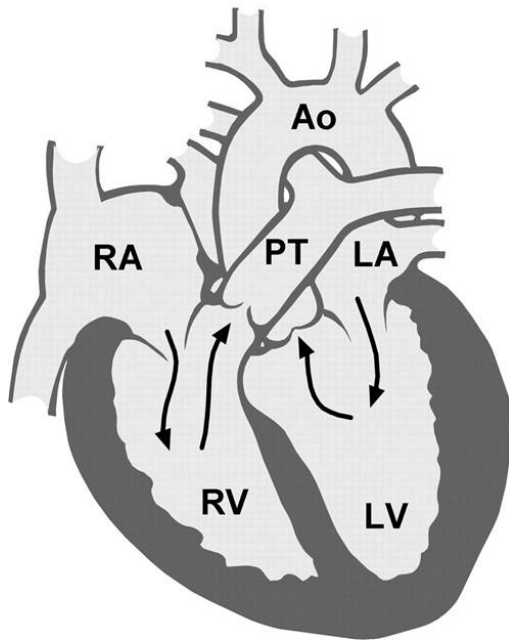


Normal



Congenitally-corrected transposition

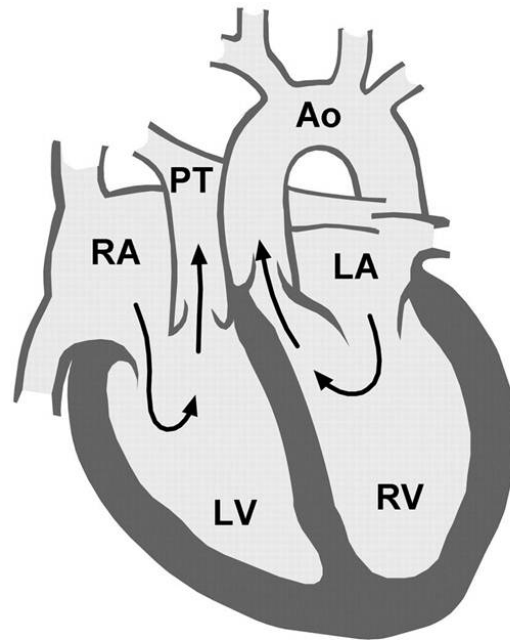


Figure 1. Anatomy of l-TGA

From Warnes CA. Transposition of the great arteries. *Circulation*. 2006;114:2699-709.

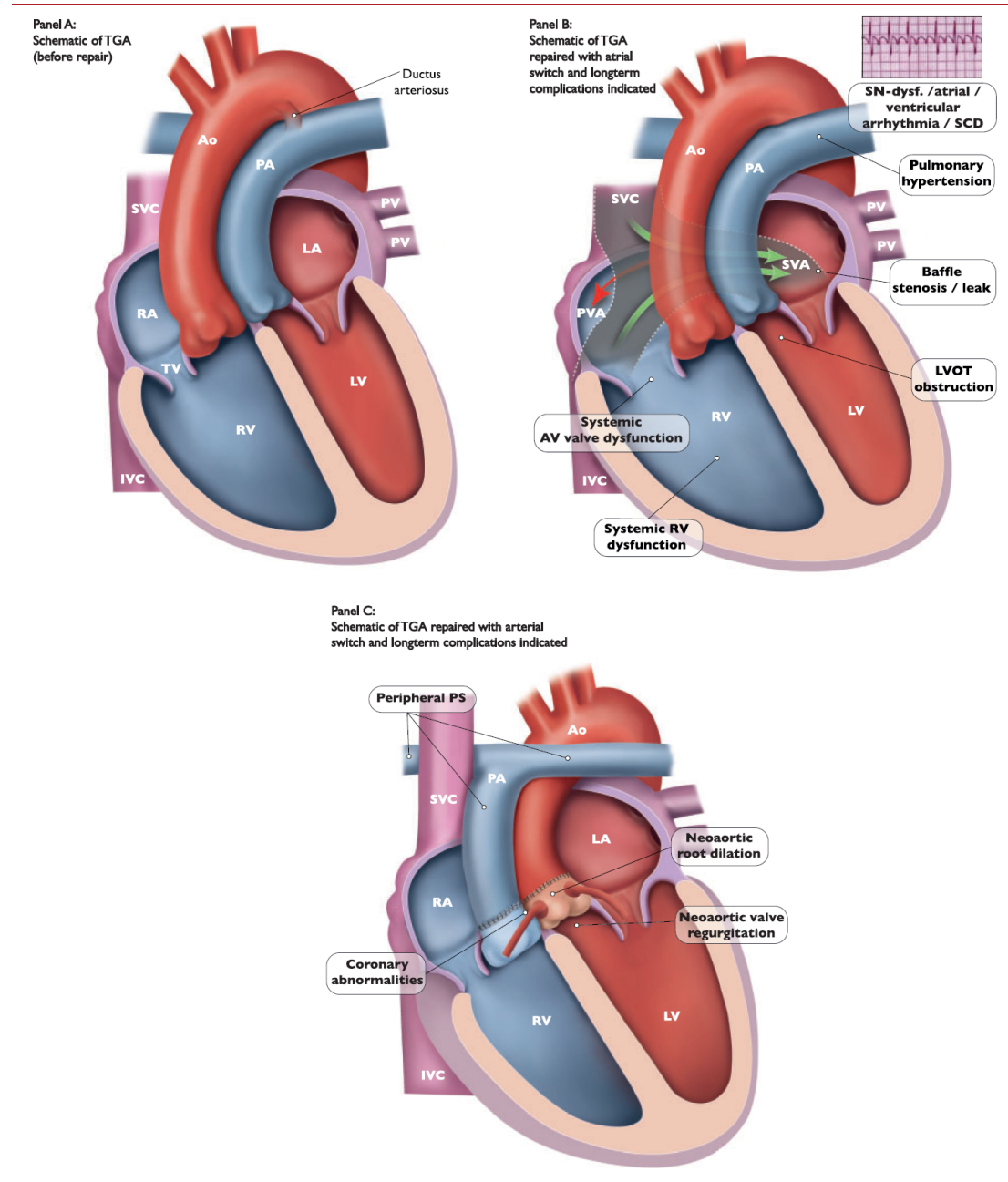


Figure 2. ESC 2020 GL on ACHD. Management of transposition of the great arteries: long-term complications to address during follow-up.

Ao = aorta; AV = atrioventricular; IVC = inferior caval vein; LA = left atrium; LV = left ventricle; LVOT = left ventricular (subpulmonary) outflow tract; PS = pulmonary stenosis (supraaortic/pulmonary artery branch); PV = pulmonary vein; PVA = pulmonary venous atrium; RA = right atrium; RV = right ventricle; SCD = sudden cardiac death; SN-dysf. = sinus node dysfunction; SVA = systemic venous atrium; SVC = superior vena cava; TV = tricuspid valve.

Table 1. ACC/AHA 2025 GL on ACHD. Recommendations for Patients with d-Transposition of the Great Arteries and Atrial Switch

Diagnostic	
Adults with dextro-transposition of the great arteries (d-TGA) and atrial switch who have progressive exercise intolerance or heart failure should undergo imaging with transthoracic echocardiogram and CMR imaging to assess ventricular, valve, baffle, and pathway function to detect and identify treatable hemodynamic lesions.	1 B-NR
Adults with d-TGA and atrial switch who present with progressive exercise intolerance or heart failure should undergo CPET to assess for potential intervention targets, including arrhythmia and desaturation.	1 B-NR
Adults with d-TGA and atrial switch with progressive exercise intolerance, heart failure, pulmonary hypertension, worsening atrial or ventricular tachyarrhythmia, pathway obstruction, or baffle leak should undergo invasive hemodynamic assessment to identify targets for medical, percutaneous, or surgical intervention.	1 B-NR
Adults with d-TGA and atrial switch who require permanent pacemaker or an ICD should undergo preprocedural imaging, in addition to intraprocedural contrast venography, to detect baffle stenosis and residual baffle leaks.	1 C-LD
For adults with d-TGA and atrial switch, biomarkers and validated disease-specific risk scores can be useful to identify patients at high risk for adverse cardiac events, prompting closer follow-up and referral to specialized heart failure transplant centers.	2a B-NR
In adults with d-TGA and atrial switch with new or progressive symptoms of heart failure, it is reasonable to obtain a resting electrocardiogram and outpatient ambulatory rhythm monitoring to ensure timely diagnosis and treatment of tachyarrhythmia or bradyarrhythmia.	2a B-NR
In adults with d-TGA and atrial switch, it may be reasonable to apply disease-specific scores to identify patients at increased risk for sustained ventricular tachyarrhythmia and/or SCD, in order to determine eligibility for a primary prevention ICD.	2b B-NR

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	12-24	12	3-6
Electrocardiogram	N/A	12-24	12	12
Transthoracic echocardiogram	N/A	12-24	12	12

Table 2. ACC/AHA 2025 GL on ACHD. Recommendations for Patients with d-Transposition of the Great Arteries and Atrial Switch

Therapeutic	
In adults with d-TGA and atrial switch who have atrial arrhythmias, rhythm control strategies are preferable to rate control strategies to reduce symptoms and prevent heart failure.	1 B-NR
Adults with d-TGA and atrial switch who have symptoms attributable to a baffle leak should undergo closure of the leak to improve symptoms and quality of life.	1 C-LD
In adults with d-TGA and atrial switch who have symptoms attributable to systemic or pulmonary venous pathway stenosis or liver congestion, intervention to relieve the stenosis is indicated to improve symptoms and prognosis.	1 C-LD
Adults with d-TGA and atrial switch who have worsening symptoms of exercise intolerance, heart failure, or arrhythmia refractory to treatment should be referred to a heart failure program for assessment for mechanical support and transplantation in consultation with an ACHD cardiologist, to improve quality of life and prolong survival.	1 C-LD
In adults with d-TGA and atrial switch who have sustained intraatrial reentrant arrhythmias or atrial fibrillation, oral anticoagulation can be useful to prevent embolic events.	2a C-LD
Adults with d-TGA and atrial switch who require transvenous lead placement can benefit from catheter-based interventions to address residual baffle leaks or obstruction before lead placement, to reduce the risk for systemic thromboembolism and/or future baffle occlusion.	2a C-EO
Symptomatic adults with d-TGA and atrial switch who have a failing systemic right ventricle and a QRS duration >150 ms related to chronic ventricular pacing may be considered for CRT to improve symptoms.	2b B-NR
In adults with d-TGA and atrial switch who have heart failure with reduced RV function, it may be reasonable to use GDMT for heart failure with reduced ejection fraction in order to treat the heart failure and improve symptoms, ventricular function, and outcome.	2b B-R

Table 3. ESC 2020 GL on ACHD. Recommendations for intervention in transposition of the great arteries after atrial switch operation

Indications for surgical intervention

Surgical repair (catheter intervention rarely possible) in symptomatic patients with pulmonary venous atrium obstruction	I C
Surgical repair in symptomatic patients with baffle stenosis not amenable to catheter intervention	I C
Surgical repair in symptomatic patients with baffle leaks not amenable to catheter-based closure	I C
In patients with severe systemic (tricuspid) AV valve regurgitation, without significant ventricular systolic dysfunction (EF >40%), valve repair or replacement regardless of symptoms.	IIa C
PA banding in adults, as LV training with subsequent arterial switch procedure, is not recommended.	III C

Indications for catheter intervention

In symptomatic patients with baffle stenosis, stenting when technically feasible.	I C
In symptomatic patients with baffle leaks and cyanosis at rest or during exercise, or with strong suspicion of paradoxical emboli, stenting (covered) or device closure when technically feasible.	I C
In patients with baffle leaks and symptoms due to L_R shunt, stenting (covered) or device closure when technically feasible.	I C
In asymptomatic patients with baffle leaks with substantial ventricular volume overload due to L_R shunt, stenting (covered) or device closure when technically feasible.	IIa C
In patients with a baffle leak who require a PM/ICD, closure of the baffle leak with a covered stent, when technically feasible, prior to insertion of transvenous leads.	IIa C
In asymptomatic patients with baffle stenosis, stenting when technically feasible.	IIb C

AV = atrioventricular; EF = ejection fraction; ICD = implantable cardioverter defibrillator; L_R = left-to-right; LV = left ventricle/ventricular; PA = pulmonary artery; PM = pacemaker.

Table 4. ACC/AHA 2025 GL on ACHD. Recommendations for Patients with d-TGA and Rastelli Repair

In adults with d-TGA and Rastelli repair, imaging is recommended to ensure timely identification of complications, including stenosis and/or regurgitation of the RV-to-PA conduit, residual VSDs, LVOT obstruction, and RV dysfunction. 1 B-NR

Patients With d-TGA and Rastelli Operation: 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	12-24	12	3-6
Electrocardiogram	N/A	12-24	12	12
Transthoracic echocardiogram	N/A	12-24	12	12

Table 5. ACC/AHA 2025 GL on ACHD. Recommendations for Arterial Switch Operation

Diagnostic

In adults with an arterial switch operation, baseline and serial imaging with both echocardiography and CMR imaging should be performed periodically to assess neo-aortic size, valve function, PA or branch PA morphology, and ventricular function. 1 B-NR

In adults with an arterial switch operation and symptoms concerning for myocardial ischemia, coronary evaluation with coronary angiography, cross-sectional imaging, and/or functional coronary assessment for an anatomic etiology should be performed. 1 C-LD

In asymptomatic adults with an arterial switch operation, baseline anatomic assessment of coronary artery anatomy and patency with gated cardiac CT is reasonable to evaluate for postoperative complications 2a B-NR

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	12-24	12	6-12	3-6
Electrocardiogram	12-24	12-24	12	6
Transthoracic echocardiogram†	12-24	12-24	12	12

CMR should include assessment of neo-aortic size, the origin and proximal course of the coronary arteries, branch pulmonary arteries, ventricular function, and valvular function.

Table 6. ACC/AHA 2025 GL on ACHD. Recommendations for Arterial Switch Operation

Therapeutic	
In adults with an arterial switch operation and evidence of myocardial ischemia, coronary revascularization should be performed to reduce symptoms and improve outcomes.	1 C-EO
In adults with an arterial switch operation and symptoms attributable to moderate or greater supraaortic or branch PA stenosis, intervention is reasonable to improve symptoms and functional class.	2a C-EO
In asymptomatic adults with an arterial switch operation who have moderate or greater supraaortic or branch PA stenosis—in addition to RV hypertension, RV dysfunction, progressive tricuspid regurgitation, and/or objective decline in exercise capacity—intervention can be useful to relieve obstruction.	2a C-EO
In adults with an arterial switch operation and severe neo-aortic root dilation who have progressive neo-aortic valve insufficiency and/or rapid root growth or for whom a neo-aortic valve intervention is planned, neo-aortic root replacement may be considered to prevent worsening neo-aortic regurgitation.	2b B-NR

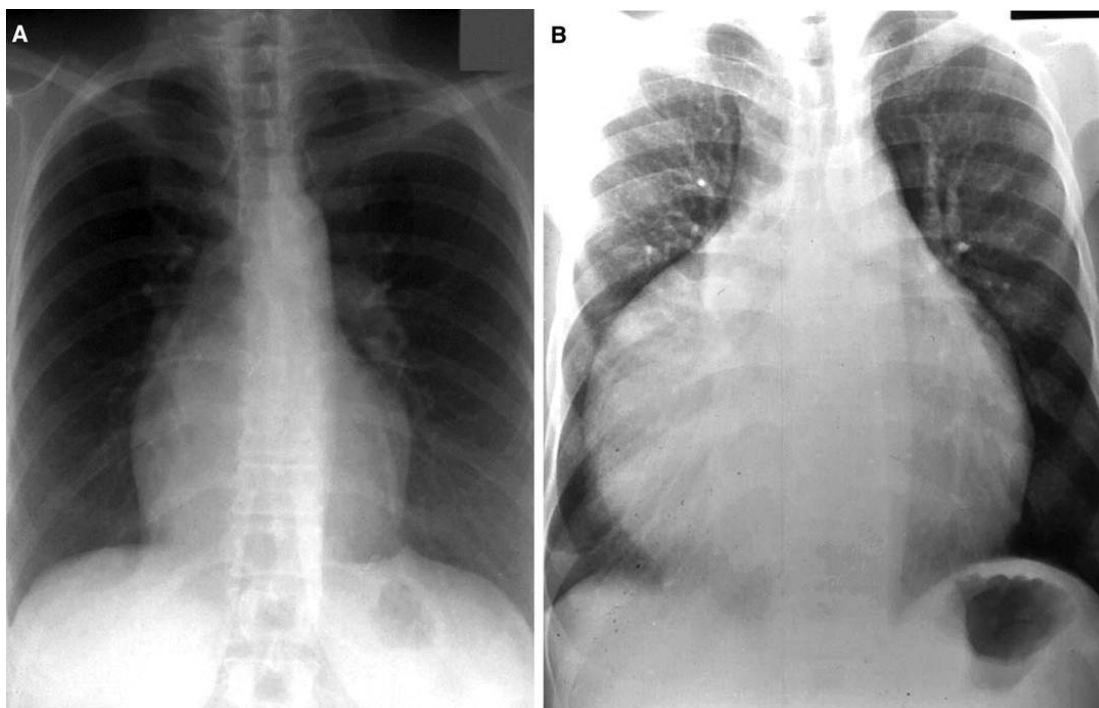


Figure 3. Chest X-rays in l-TGA

From Warnes CA. Transposition of the great arteries. *Circulation*. 2006;114:2699-709.

Table 7. ACC/AHA 2025 GL on ACHD. Recommendations for Congenitally Corrected Transposition of the Great Arteries

Diagnostic	
In adults with congenital CCTGA, periodic* transthoracic echocardiography is recommended to assess chamber function, systemic tricuspid valve function, and associated anatomic lesions such as septal defects and LVOT stenosis.	1 B-NR
In adults with CCTGA, periodic cross-sectional imaging (preferably, CMR) is reasonable to assess systemic ventricular volumes and function, quantify the severity of systemic tricuspid regurgitation, and assess the severity of associated anatomic lesions.	2a B-NR
In adults with CCTGA without symptoms, periodic ambulatory rhythm monitoring is reasonable to screen for high-grade atrioventricular block.	2a B-NR

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	12	6
Electrocardiogram	N/A	12	12	12
Transthoracic echocardiogram	N/A	12	12	12

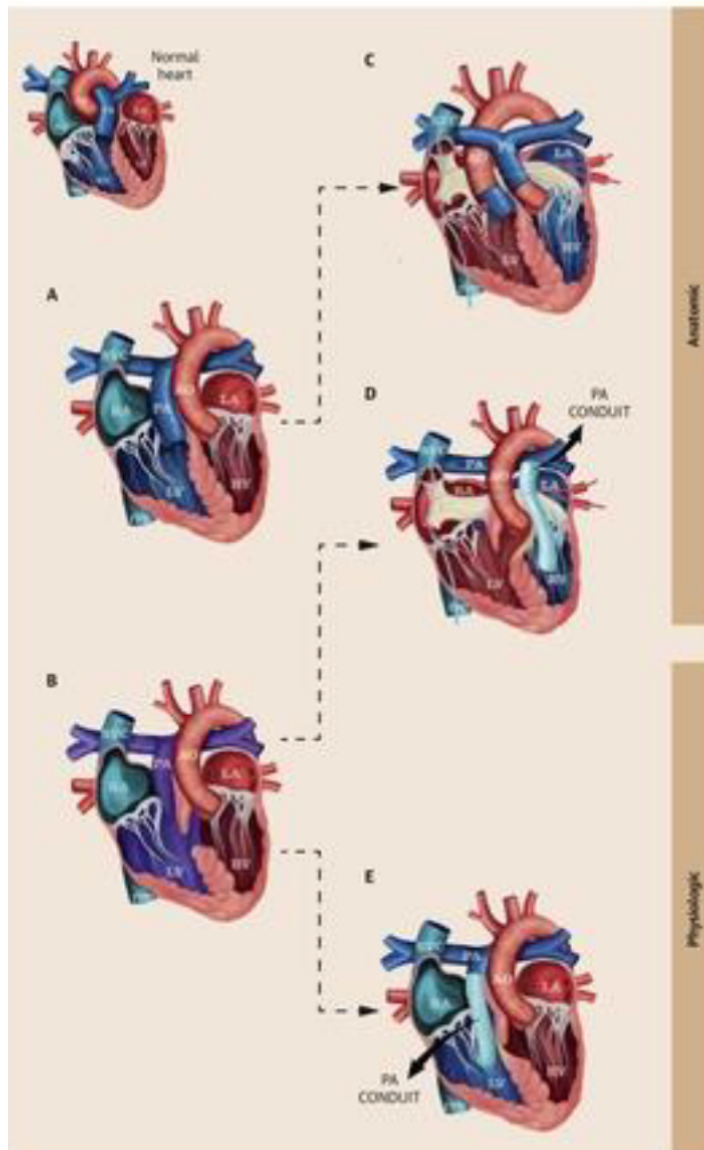


Figure 4. Anatomic and Physiologic Repair in Patients With Congenitally Corrected Transposition of the Great Arteries

The illustration shows 2 phenotypes of ccTGA in levocardia with different repair techniques. The drawing on the top left represents a normal heart. (A) ccTGA. (B) ccTGA with VSD and pulmonic stenosis. (C) Double switch operation. (D) Rastelli-Mustard operation with VSD closure. (E) LV-PA

conduit with VSD closure. The graphs show the meta-analysis depicting the risk of (left) mortality/heart transplantation and (right) reintervention in anatomic and physiologic repair across the different follow-up periods.

AO = aorta; ccTGA = congenitally corrected transposition of the great arteries; IVC = inferior vena cava; LA = left atrium; LV = left ventricle; n = number of events; N = sample size; PA = pulmonary artery; P(Het) = *P* value for heterogeneity; RA = right atrium; RV = right ventricle; SVC = superior vena cava; VSD = ventricular septal defect.

Jacob KA, et al. Anatomic and Physiologic Repair of Congenitally Corrected Transposition of the Great Arteries. *Journal of the American College of Cardiology*. 2024;84:2471-2486.

Table 8. ACC/AHA 2025 GL on ACHD. Recommendations for Congenitally Corrected Transposition of the Great Arteries

Therapeutic	
In symptomatic adults with CCTGA, severe systemic tricuspid regurgitation, and normal or mildly impaired RV systolic function, surgical tricuspid valve replacement is recommended to relieve symptoms and preserve RV function.	1 B-NR
In asymptomatic adults with CCTGA, severe systemic tricuspid regurgitation, and normal or mildly impaired RV systolic function, surgical tricuspid valve replacement is reasonable to preserve RV systolic function.	2a B-NR
In symptomatic adults with CCTGA and high-grade atrioventricular block, physiological pacing (CRT or conduction system pacing) is reasonable to relieve symptoms.	2a B-NR
In adults with CCTGA and heart failure with RV dysfunction, the use of GDMT may be reasonable to improve ventricular function and functional capacity.	2B B-NR
In symptomatic adults with CCTGA and severe LVOT stenosis, surgical or transcatheter therapy to relieve LVOT obstruction may be considered to improve symptoms and preserve LV systolic function while accounting for the potential risk of worsening the severity of tricuspid regurgitation. ¹³	2b B-NR

Table 9. ESC 2020 GL on ACHD. Recommendations for intervention in congenitally corrected transposition of the great arteries

TV replacement in symptomatic patients with severe TR and preserved or mildly impaired systemic RV systolic function (EF >40%)	I C
TV replacement in asymptomatic patients with severe TR and progressive systemic RV dilatation and/or mildly impaired systemic RV systolic function (EF >40%).	IIa C
Biventricular pacing should be considered in case of complete AV block or >40% ventricular pacing requirement.	IIa C

In symptomatic patients with severe TR and more than mildly reduced systemic RV systolic function (EF < 40%), TV replacement may be considered.

IIB C

AV = atrioventricular; EF = ejection fraction; RV = right ventricle/ventricular; TR = tricuspid regurgitation; TV = tricuspid valve.

Table 10. ESC 2025 GL on Pregnancy.

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
Transposition of the great arteries				
TGA after atrial switch (Mustard or Senning) and CCTGA	- High pregnancy risk—MACE rate up to 28% in retrospective series⁴⁷⁰—lower risk in ROPAC⁶. Atrial and ventricular arrhythmias in 6.7%, HF in 10% - Atrial arrhythmias often poorly tolerated - Baffle leaks may lead to desaturation and paradoxical embolism⁴⁷¹ - Predictors of MACE: symptoms of HF before pregnancy and systemic RV EF <40%⁶	- Foetal risks associated with maternal CO and saturation - Pre-term birth (21%) - Low birth weight (18%–21%)^{6,472,473} - Rare foetal and neonatal death (1%)⁴⁷⁴ - PPH (7%)⁶	- According to anatomical and functional status: TTE every 1–3 months and serial NP - Holter monitoring if palpitations	- Pre-pregnancy counselling about very high risk if NYHA class III/IV, systemic RV EF <40%, more than moderate tricuspid regurgitation, or treated HF^{472,473,475} - Treat HF primarily with medical therapy^h - Promptly treat arrhythmia^g - Consider prolonged post-partum monitoring (48–72 h) and early post-partum FU given the ↑ risk of post-partum HF No clear evidence for long-term deterioration or ↑ cardiovascular events associated with pregnancy^{6,476}
			regurgitation and ↑ aortic dilatation	