

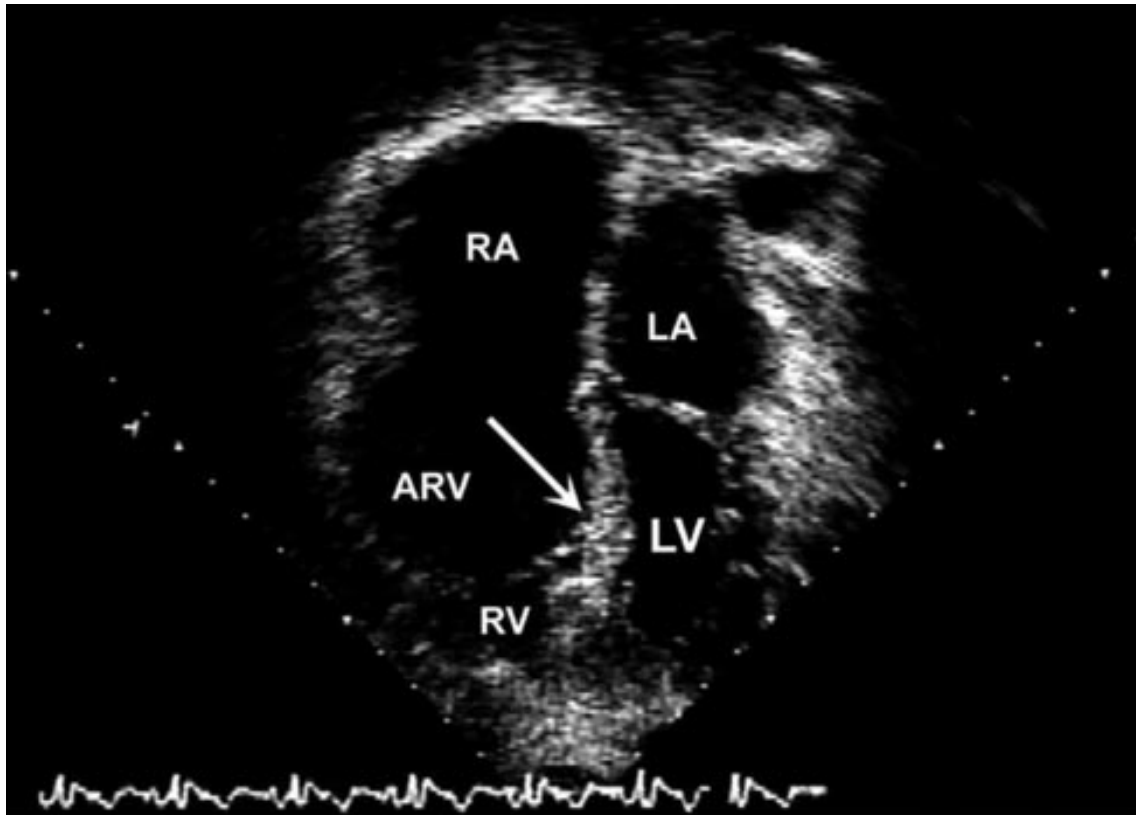


Figure 1. Ebstein's anomaly.

Example of an echocardiogram (4-chamber view, apex down) of a patient with severe Ebstein's anomaly showing a grossly displaced septal leaflet (arrow). The anterior leaflet is severely tethered and nearly immobile. The functional right ventricle (RV) is small.

ARV indicates atrialized right ventricle; LA, left atrium; LV, left ventricle; and RA, right atrium.

From Attenhofer Jost CH, Connolly HM, Dearani JA, et al. Ebstein's anomaly. *Circulation*. 2007;115:277-85.

Table 1. AHA/ACC 2025 GL on ACHD. Recommendations for Ebstein Anomaly

Diagnostic	
In adults with Ebstein anomaly, an electrophysiological study is recommended for patients with symptomatic arrhythmia or asymptomatic ventricular preexcitation to assess for substrates for ablation, including multiple or high-risk accessory pathways.	1 B-NR
In adults with Ebstein anomaly, CMR imaging can be useful to determine anatomy, RV dimensions, and systolic function to guide surgical planning and management.	2a B-NR
In adults with Ebstein anomaly, electrophysiological study (and catheter ablation, if needed) can be useful before tricuspid valve surgery to improve clinical outcomes.	2a B-NR
In adults with Ebstein anomaly, if transthoracic echocardiography is inadequate to guide surgical planning, transesophageal echocardiography can be beneficial to characterize tricuspid valve morphology and function.	2a C-LD
In adults with Ebstein anomaly and severe RV dysfunction in whom there is concern that the RV will not support the entire stroke volume, preoperative diagnostic cardiac catheterization may be considered to assess candidacy for a bidirectional nsuperior cavopulmonary (Glenn) anastomosis	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	12-24	12	6-12	3-6
Electrocardiogram	12-24	12	12	12
Transthoracic echocardiogram†	12-24	12-24	12	12

†Routine transthoracic echocardiogram may be unnecessary in a year when CMR imaging is performed, unless clinical indications warrant otherwise.

Therapeutic

In adults with Ebstein anomaly and at least moderate tricuspid regurgitation in the presence of heart failure, worsening exercise capacity, or progressive RV systolic dysfunction, surgical intervention is recommended to restore valve function and augment cardiac output.	1 B-NR
In adults with Ebstein anomaly and high-risk accessory pathway conduction or multiple accessory pathways, catheter ablation is recommended to reduce the risk for SCD.	1 B-NR
In asymptomatic adults with Ebstein anomaly and at least moderate tricuspid regurgitation in the presence of progressive RV enlargement, systemic desaturation from a right-to-left atrial shunt, recurrent arrhythmias, and/or paradoxical embolism, surgical intervention can be beneficial to improve clinical outcomes.	2a B-NR
In adults with Ebstein anomaly and severe RV dysfunction in whom there is concern that the RV will not tolerate supporting the entire stroke volume, a bidirectional superior cavopulmonary (Glenn) anastomosis at the time of tricuspid valve surgery may be considered in carefully selected individuals to improve cardiac output through ventricular unloading.	2b B-NR
In adults with Ebstein anomaly who have an ASD/patent foramen ovale, and cyanosis or paradoxical embolism, isolated closure may be considered after hemodynamic assessment to improve clinical outcomes.	2b C-LD

Table 3. ESC 2020 GL on ACHD. Recommendations for intervention in Ebstein anomaly

Indications for surgery

Surgical repair in patients with severe TR and symptoms or objective Deterioration of exercise capacity. I C

Surgical repair to be performed by a congenital surgeon with specific experience in Ebstein surgery. I C

If there is an indication for TV surgery, ASD/PFO closure at the time of valve repair if it is expected to be haemodynamically tolerated. I C

Surgical repair regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function. IIa C

Indications for catheter intervention

In patients with symptomatic arrhythmias, or pre-excitation on the ECG, Electrophysiologic testing followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery. I C

In the case of documented systemic embolism, probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered but requires careful evaluation before intervention to exclude induction of RA pressure increase or fall in cardiac output. IIa C

If cyanosis (oxygen saturation at rest <90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention to exclude induction of RA pressure increase or fall in cardiac output. IIb C

ASD = atrial septal defect; ECG = electrocardiogram; PFO = patent foramen ovale; RA = right atrium/atrial; RV = right ventricle/ventricular; TR = tricuspid regurgitation; TV = tricuspid valve.

Table 4. ESC 2025 GL on Pregnancy

ACHD	Maternal Risk	Obstetric and foetal risk	Monitoring	Pregnancy management and delivery
Ebstein anomaly				
	- Overall MACE rate in ROPAC 9.9% - Low risk in mild/ moderate Ebstein (Very) high risk when pre-pregnancy HF, cyanosis due to atrial shunt ↑ Arrhythmia risk due to accessory pathways⁴⁶⁹	- Foetal risk is related to ↓ maternal CO and cyanosis^{3,469} - Miscarriage - Pre-term birth (20%–24%) - Neonatal death (3%) - PPH - Recurrence risk (5%)	<i>Mild to moderate:</i> baseline and 28–32 weeks assessment with TTE and ECG <i>Severe:</i> monthly/ bimonthly TTE & ECG - Monitor for arrhythmias if palpitations	- Severe tricuspid regurgitation with HF can usually be managed medically - Treat arrhythmias promptly⁸ - Appropriate pre-pregnancy counselling about very high risk of MACE when HF and/or cyanosis