

2020 ESC Guidelines for: Management of Adult Congenital Heart Disease (ACHD)

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Introduction

These guidelines provide a significant update in the structuring of care, assessment and management of the broad spectrum of pathologies encompassing ACHD. There are no comprehensive national guidelines and UK practice is informed by international guidelines and expert opinion within regional multidisciplinary teams.

Common themes

Heart Failure

Heart failure is an increasingly prevalent presentation within the ACHD population. The pathophysiology is often distinct from 'acquired' heart disease and there is a paucity of evidence for the effectiveness of conventional therapies. In addition to cautious medical therapy, great emphasis is placed on correcting specific haemodynamic triggers including arrhythmias and adverse structural heart defects.

Conventional medical therapies are advised in the impaired systemic left ventricle with a biventricular circulation and can be considered in the symptomatic impaired systemic right ventricle. No clinical benefit has been demonstrated for the impaired sub-pulmonary ventricle (e.g. right ventricle in Tetralogy of Fallot (ToF)), asymptomatic systemic right ventricle (e.g. congenitally corrected transposition of the great arteries (ccTGA) or post-atrial switch repair for transposition of the great arteries (TGA)) or single ventricle physiology (e.g. Fontan circulation). Furthermore, unconsidered use of such therapies may be harmful by adversely affecting the precarious balance of ventricular preload and systemic afterload.

The increasing use of advanced therapies including inotropes, ventricular assist devices and ECMO is acknowledged but guidance is not offered in patient selection other than early referral and assessment in experienced centres.

Arrhythmia and Pacing

Emphasis is placed on the prompt identification and treatment of arrhythmias in pursuit of preserving sinus rhythm and prevention of ventricular arrhythmias. There is a significant and differing burden of arrhythmia substrates related to each condition and type of repair. Many circulations poorly tolerate arrhythmia and have an increased risk of sudden cardiac death (SCD) associated with atrial and re-entrant tachycardias. Therefore, reversible causes should be treated

aggressively and moderate or severe complexity ACHD referred to experienced ACHD centres (Class I). Catheter ablation is recommended for symptomatic and recurrent atrial or re-entrant tachycardias in preference to medical therapy (Class I/IIA, C) and considered adjunctive in ventricular arrhythmia (Class IIA, C).

Bradycardia is recognised to promote intra-atrial re-entrant tachycardia which can rapidly decompensate circulations dependent on effective atrio-ventricular synchrony. In these patients, broader pacing indications for sinus node disease are recommended (daytime bradycardia <40bpm or pauses >3s) (Class IIA, C).

Conventional indications for defibrillator (ICD) and resynchronisation therapy (CRT) are applicable to ACHD conditions with a systemic left ventricle. Primary prevention ICD can be considered in severely impaired single ventricle or systemic right ventricular physiology (Class IIB, C) as well as ToF (Class IIA, C) with recognised risk factors for SCD. The complexities of devices implantation in these circulations however must not be underestimated and should be carried out in centres with expertise. The broader application of CRT is unclear and QRS duration may not be a sufficient predictor of benefit. It is considered reasonable in patients with a severely impaired systemic ventricle and narrow QRS who have an anticipated significant pacing requirement.

Catheter interventions

Accumulating evidence advocates the safety and efficacy of catheter intervention for atrial septal defects, coarctation and right ventricular outflow tract obstruction (RVOTO). These offer an attractive alternative to surgical intervention and are the recommended treatment, if feasible, in experienced ACHD centres.

Prior to closure of septal defects, care in assessment of pulmonary vascular resistance (PVR) is advised with invasive PVR testing mandated in patients with non-invasive signs of raised pulmonary artery pressure (PAP). Closure is recommended in shunts with raised pulmonary flow ($Q_p:Q_s > 1.5$) and low PVR (<3WU) (Class I, C) and suggested in patients with moderately raised PVR (3-5WU) (Class IIA, C). Fenestrated atrial septal defect (ASD) closure can be considered in selected patients with raised PVR (≥ 5 WU) who respond to targeted pulmonary arterial hypertensive therapy (Class IIB, C). Caution is advised in patients with impaired systemic ventricular function as ASD closure can raise systemic filling pressures and worsen heart failure.

Haemodynamic confirmation of aortic coarctation is recommended prior to intervention (peak to peak differential of ≥ 20 mmHg) (Class I, C). Finally, percutaneous pulmonary valve implantation to relieve RVOTO is now the recommended intervention, when feasible, in non-native outflow tracts (Class I, C).

There is no guidance on the role of trans-catheter aortic valve implantation in left ventricular outflow tract obstruction (LVOTO).

Key guidance on specific conditions

- **LVOTO:** Surgical intervention is recommended in symptomatic obstruction, at any level, with a lower mean gradient threshold of ≥ 40 mmHg (Class I, C). In sub- and supra- valvular stenosis it can be considered in asymptomatic patients with normal systemic ventricular

function without exercise test abnormalities if there is low surgical risk (Class IIB, C). Intervention can also be considered in valvular stenosis within these parameters with a raised BNP ($\times 3$ normal) or related severe pulmonary hypertension (invasive PAP >60 mmHg) (Class IIA, C).

- RVOT dysfunction (ToF and RV-PA conduits): Specific right ventricular volumetric criteria are suggested to guide timing of intervention in asymptomatic severe pulmonary regurgitation ($RVEDVi \geq 160$ ml/m²). Sustained arrhythmias alone are no longer considered an indication for intervention.
- TGA/ccTGA: Identification and closure of baffle leaks are recommended before trans-venous device implantation (Class IIA, C).
- Univentricular heart: All patients with these palliated or un-operated circulations should undergo assessment with multimodal imaging and invasive haemodynamic testing in ACHD centres to determine whether they would benefit from structural interventions (Class I, C).
- Fontan circulation: Sustained atrial arrhythmias with rapid ventricular conduction are a medical emergency to be treated with synchronised cardioversion (Class I, C). Anticoagulation is recommended for intra-atrial re-entrant tachycardia or atrial fibrillation regardless of conventional risk scoring. Additionally, a low threshold for invasive haemodynamic assessment is recommended in unexplained deterioration (oedema, reduced exercise tolerance, arrhythmias, cyanosis or haemoptysis) (Class I, C). Whilst regular imaging for Fontan associated liver disease (FALD) is recommended, no specific guidance is offered in the management of FALD, plastic bronchitis or protein losing enteropathy.
- Aortopathy: Valve repair with coronary re-implantation is recommended in young patients meeting threshold criteria of annular dilatation and tricuspid aortic valves (Class I, C). There is significant expansion of guidance in line with the 2014 ESC aortopathy guidelines.
- Coronary artery abnormalities: Functional imaging for ischemia is recommended all patients (Class I, C). Surgical intervention is recommended in anomalous right coronary artery from the pulmonary artery with symptoms (Class I, C) or ischemia (Class IIA, C) and all patients with anomalous left coronary artery from the pulmonary artery (Class I, C). Guidance is presented for risk stratification of anomalous aortic origin of coronary arteries dependent on symptoms, ischemia, anatomy and age.
- ACHD with pulmonary hypertension: Emphasis is placed on differentiating pre-capillary pulmonary hypertension from post-capillary hypertension in line with the 2015 ESC pulmonary hypertension guidelines. The former group are most likely to benefit from targeted pulmonary arterial hypertensive treatments and should be risk assessed in an expert centre (Class I, C) and advised against pregnancy (Class I, C). Direct measurement with right heart catheterisation is advised prior to major decision making including commencement of targeted therapies, pregnancy or surgery. Those deemed low or intermediate risk should have initial sequential targeted therapy (or combination) with high risk patients advised to have initial combination therapy including parenteral prostanoids (Class I, A).
- Eisenmenger's syndrome: Reduced exercised capacity (6MWT <450 m) should be treated proactively with initial endothelin receptor antagonists followed by combination pulmonary arterial hypertension therapies if needed (Class IIA, B). Supplemental oxygen should only be administered if it produces a consistent and significant increase in oxygen saturations with symptom improvement.

Conclusions

This guideline presents a significant expansion and update in the management of the spectrum of congenital heart disease. All patients with ACHD are recommended to be reviewed at least once by a specialist centre to determine their suitability for intervention, management of complications and appropriate follow up.