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Management Guidelines for Adults with Congenital Heart Disease

Royal Brompton Hospital Congenital Heart Disease Network

These are the agreed recommendations from the Royal Brompton Hospital Adult Congenital Heart Disease Consultant Group, the Congenital Heart Disease Operational Delivery Network Board and the Congenital Heart Disease Operational Delivery Network Pathways & Protocols Sub-Committee

These guidelines form part of the Congenital Heart Disease Operational Delivery Network Standard Operating Procedure

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RBH has adapted American Heart Association Guidance for management of Adult CHD patients.

2 Physiological States

A

- NYHA FC I symptoms
- No hemodynamic or anatomic sequelae
- No arrhythmias
- Normal exercise capacity
- Normal renal/hepatic/pulmonary function

B

- NYHA FC II symptoms
- Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular
- enlargement, mild ventricular dysfunction)
- Mild valvular disease
- Trivial or small shunt (not hemodynamically significant)
- Arrhythmia not requiring treatment
- Abnormal objective cardiac limitation to exercise

C

- NYHA FC III symptoms
- Significant (moderate or greater) valvular disease; moderate or greater
- ventricular dysfunction (systemic, pulmonic, or both)
- Moderate aortic enlargement
- Venous or arterial stenosis
- Mild or moderate hypoxemia/cyanosis
- Hemodynamically significant shunt
- Arrhythmias controlled with treatment
- Pulmonary hypertension (less than severe)
- End-organ dysfunction responsive to therapy

D

- NYHA FC IV symptoms
- Severe aortic enlargement
- Arrhythmias refractory to treatment
- Severe hypoxemia (almost always associated with cyanosis)
- Severe pulmonary hypertension
- Eisenmenger syndrome
- Refractory end-organ dysfunction

3 Atrial Septal Defect

Confirm Diagnosis

- Referral to Level 1 centre
- Type of ASD and size confirmed via TTE ECHO

Assessment

- Chest X-ray; Blood test, ECG; CMR; MVO2;
- TOE ECHO might be necessary to assess type and size of ASD and asscoaited defects (venous drainage)
- Cardiac Catheter may be required to assess for pulmonary vacsular resistence, LV diastolic dysfunction and any other associated defects.

Intervention

- **Device closure** is first choice intervention for ASD OS if size and rims are adequate.
- Antiplatelet therapy recommended for at least 6 months post percutaneous intervention.
- **Surgical intervention** is recommended for : ASD OS not suitable for percutaneous closure; primum ASD; and most sinous venous ASDs.

Post intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks to assess the device position, pericardial effusion; residual shunt; TR; PAP.
- ECG to ensure sinus rhythm. If concerned about arrhythmia holter monitor should be ordered

Long-term Follow-up

- Patients with significant residual shunts; elevated PAP; or arrhythmias before or after repair should be reviewed at a level 1 centre at least once every 1-2 years.
- Patients with Eisenmenger syndrome or PH associated with closed defects should have specialist PHT follow-up every 3 - 12 months depending on PHT therapies and severity (refer to PH4 clinic).

4 Ventricular Septal Defect

Confirm Diagnosis

- Referral to Level 1 Centre
- Type of VSD and size confirmed via TTE ECHO.

Assessment

- Chest X-ray; Bloods, ECG; CMR; MVO2;
- RH Cardiac Catheter may be required to assess PVR, Qp/Qs and any other associated defects.

Intervention

- Closure should be considered:
  - If unrepaired VSD with significant shunt (Qp/Qs >1.5) and PVR not elevated
  - If restrictive VSD but progressive AR, LV dilatation or history of endocarditis
- **Surgical closure** remains the treatment of choice in most cases.
- **Device closure** can be considered when the defect is accessible for percutaneous intervention or in patients with increased risk factors for surgery.

Post Intervention Follow-up

- Post procedure CNS Review with ECHO within 6-8 weeks to assess for pericardial effusion, residual shunt, TR and estimated PAPs.
- ECG to ensure sinus rhythm. If concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Patients with LV dysfunction; residual shunt; AR; RVOTO; LVOTO should be seen every 1 year at Level 1 centre
- Patients with small VSD (native or residual) and normal LV; normal PAP; asymptomatic; and no other lesion should be reviewed every 3 -5 years.
- After device closure annual follow-up for the first 2 years then if no complications follow-up should be every 2-4 years
- After surgical closure without residual abnormalities follow-up should be every 3-5 years.
- Patients with Eisenmenger syndrome or PH associated with closed defects should have specialist PHT follow-up every 3 - 12 months depending on PHT therapies and severity (refer to PH4 clinic).
- If residual shunt after closure OR Eisenemenger Syndrome --> Endocarditis Prophylaxis for invasive dental procedures is recommended

5 Atrioventricular Septal Defect

Confirm Diagnosis

- Referral to Level 1 Centre
- Diagnosis confirmed via TTE ECHO:
  - - **Complete AVSD** is characterized by the presence of a common AV annulus guarded by 5 leaflets; the septal defect is in the crux of the heart, extending to both the intral atrial and intraventircular septum.
  - - **Partial AVSD or Primum ASD** - the anterior and posterior bridging leaflets are fused centrally creating seperate left and right-sided orifices; the septal defect is only at atrial level.
  - - **Transional or intermediate AVSD** has an incomplete atrial and VSDs and/or incomplete abnormalities of the common atrioventricular valve

Assessment

- Chest X-ray; ECG; Bloods, CMR; MVO2;
- RH Cardiac Catheter may be required to assess for PHT and any other associated defects such as Eisenmenger syndrome.

Intervention

- Primary surgical repair is recommended if left-to-right shunt (Qp:Qs ≥1.5:1), PA systolic pressure less than 50% systemic and pulmonary vascular resistance less than 1/3 systemic.
- If Eisenmenger Syndrome Physiology present then intervention is contraindicated.
- Operation for discrete LVOT obstruction in adults with atrioventricular septal defect is reasonable with a maximum gradient ≥50 mm Hg a lesser gradient if HF symptoms are present, or if concomitant moderate-to-severe mitral or AR are present.
- Severe LAVV stenosis or regurgitation should bve followed as per the VHD guidelines

Post Intervention Follow-up

- Post proedure ECHO within 6-8 weeks to assess for pericardial effusion; residual shunt; TR; PAP.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Patients with residual shunt; AV malfunction; LV & RV enlargement and dysfunction; PAP elevation; LVOTO; and arrhythmias/conduction disease hould have follow-up every 1-2 years depending on the severity of the abnormality.
- Patients with no residual abnormalities and no AV valve malformation should be reviewed every 2-3 years.
- Holter monitor should be considered as complete heart block can appear as late as 15 years after surgery.
- Patients with Eisenmenger syndrome should have specialist PHT follow-up every 3 - 12 months depending on PHT therapies and severity.

6 Patent Ductus Arteriosus

Confirm Diagnosis

- Referral to Level 1 Centre
- Diagnosis confirmed via ECHO showing size of the defect, direction of shunt and LV volume overload.

Assessment

- Chest X-ray; ECG; Bloods. CMR; MVO2; Cardiac Catheter may be required to assess PVR
- Small and Moderate size PDA (but not tiny) usually cause significant left-to-right shunt, LV overload and increased pulmonary pressures.
- Unrepaired large PDA patients will likely have developed Eisenmenger Syndrome when they arrive to adulthood.

Intervention

- Device closure is the intervention of choice
- Surgery is reserved for patients where the duct is too large for device closure or with unsuitable anatomy with aneurysm formation.
- If Eisenmenger Syndrome present then intervention is contraindicated

Post Intervention Follow-up

- Post procedure CNS Review with ECHO within 6-8 weeks to assess device position, LV size and function; PAP; residual shunt; and associated lesions.
- Endocardiis prophylaxis on the first 6 months after closure.

Long-term Follow-up

- **Patients with no residual shunt,** normal LV and normal PAP should have follow-up 6 -12 months after intervention with an echocardiogram.
- **If surgical closure and no abnormality found**, patients can be discharged from specialist follow-up. If device closure, patients should be seen every 3-5 years.
- Patients with LV dysfunction or any other associated abnormalities should have follow-up every 1-3 years depending on the severity.
- **Patients with Eisenmenger syndrome or PH associated with closed defect** should have specialist PHT follow-up every 3 - 12 months depending on PHT therapies and severity.

7 Congenital Valvular Aortic Stenosis

Confirm Diagnosis

- Referral to Level 1 Centre

Assessment

- Chest X-ray; ECG; Bloods, CMR; MVO2 (except if symptomatic AS);
- Patients with bicuspid aortic valve should be screened for Aorthopathy and Aortic Coartation.
- Stress ECHO may be required to assess severity of AS and associated AR

Intervention

- **Balloon valvuloplasty** may be considered only in selected adolescents and young adults with non-calcified valves and no more than mild AR.
- **Valve Replacement** is the treatment of choice in patients with calcified valves - with mechanical or biological valves.
- **Aortic root or ascending aorta replacement** should be cnsidered when are > 45mm
- **The Ross Procedure** can be considered in those patients of child-bearing age where anticoagulation would be avoided.
- **Transcatheter valve implantation** should only be considered in those unsuitable for conventional surgery.

Post Intervention Follow-up

- Post procedure CNS review with ECHO within 6-8 weeks to assess the AVR, residual AR, root/ascending aorta, LV function; LVH and associated lesions.
- ECG to ensure sinus rhythm. If concerned about arrhythmia holter monitor should be ordered.
- Bloods to check Hb, Iron studies and inflammatory parameters.

Long-term Follow-up

- **At risk Stage A - with Vmax <2m/s** - OPA with ACHD Team with ECG and ECHO every 36-60 months (Exercise test as required)
- **Progressive Stage B** - OPA with ACHD Team with ECG and ECHO- mild severity Vmax 2.0-2.9 m/s - every 36-60 months; moderate severity Vmax 3.0-3.9m/s - every 12-24 months), (Exercise test as required).
- **Severe Stage C** - OPA with ACHD Team with ECG and ECHO every 6-12 months (Exercise test as required).
- **Aortic dilation >4.5cm** OPA with ACHD Team with ECG and ECHO every 12 months (Exercise test, CMR or CT as required).

- It is reasonable to screen first degree relatives of patients with bicuspid aortic or unicuspid aortic valve with ECHO for valve disease and aortopathy.

8 Subaortic Stenosis

Confirm Diagnosis

- Referral to Level 1 Centre
- SubAS may be first diagnosed in adulthood and high level of suspicion is required, as it may be confused with hypertrophic obstructive cardiomyopathy when LV hypertrophy of sufficient severity has developed such that subAo membrane is less evident on imaging.

Assessment

- Chest X-ray; ECG; Echo, Bloods, CMR;
- Special attention needs to be given to the degree of AR
- Cardiac Catheterization may be required to assess LVOT gradient and any other associated defects. If ischemic symptoms, coronary artery disease need to be rule out.
- Stress testing for adults with LVOT obstruction to determine exercise capacity, symptoms, ECG changes, or arrhythmias may be reasonable in the presence of otherwise equivocal indications for intervention.

Intervention

- **Surgical intervention** is recommended for:
  - Adults with subAS with a peak gradient ≥50mmHg and HF or ischaemic symptoms, and/or LV systolic dysfunction attributable to subAS.
  - Peak gradient <50mmHg and HF or ischaemic symptoms, and/or LV systolic attributable to subAS.
  - Surgery can be considered in asymptomatic adults with subAS and at least mild AR and peak gradient ≥50mmHg to prevent progression of AR

Post Intervention Follow-up

- Post procedure CNS review with ECHO within 6-8 weeks to assess residual AR, residual LVOT, LV function; LVH and associated lesions.
- ECG to ensure sinus rhythm. If concerned about arrhythmia holter monitor should be ordered.
- Bloods to check Hb, Iron studies and inflammatory parameters.

Long-term Follow-up

- Physiological Stage A - OPA with ACHD Team with ECG and ECHO every 24 months (Exercise test as required)
- Physiological Stage B - OPA with ACHD Team with ECG and ECHO every 24 months (Exercise test as required)
- Physiological Stage C - OPA with ACHD Team every 6-12 months with ECG and ECHO every 12 months. Exercise test every 24 months.
- Physiological Stage D - OPA with ACHD Team every 3-6 months with ECG and ECHO every 12 months. Exercise test every 12 months.

9 Supravalvular Aortic Stenosis

Confirm Diagnosis

- Referral to Level 1 centre
- Supravalvular aortic stenosis is a relatively rare condition overall but is seen commonly in patients with Williams syndrome or homozygous familial hypercholesterolemia.
- The stenotic ridge tends to occur distal to the coronary artery orifices at the sinotubular junction.
- Patients with supravalvular AS have higher risk of CAD and SCD.

Assessment

- ECG, bloods, Chest Xray, ECHO.
- Aortic imaging with with TOE, CMR, CT angiography is recommended in adults with Williams syndrome or patients suspected of having supravalvular AS.
- Coronary imaging is recommended in patients with Williams syndrome and supravalvular AS presenting with symptoms of coronary ischaemia

Intervention

- **Surgical repair** is recommended for adults with supravalvular AS (discrete or diffuse) and symptoms or decreased LV systolic function deemed secondary to aortic obstruction.
- **Coronary artery revascularisation** is recommended in symptomatic aduts with supravalvular AS and coronary ostial stenosis.

Post intervention Follow-up

- Post procedure CNS Review with ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered
- Bloods to assess Hb, iron studies and inflammatory parameters.

Long-term Follow-up

- Physiological Stage A - OPA with ACHD Team with ECG and ECHO **every 24 months, CMR every 36-60 month**s (Exercise Test as required).
- Physiological Stage B - OPA with ACHD Team with ECG and ECHO **every 24 months, CMR every 36-60 months**, Exercise test every 24 months.
- Physiological Stage C - OPA with ACHD Team every **6-12 months with ECG and ECHO every 12 months. CMR every 36-60 months**, Exercise test every 24 months.
- Physiological Stage D - OPA with ACHD Team every **3-6 months with ECG and ECHO every 12 months, CMR every 36-60 months**, Exercise test every 12 months.

10 Coarctation of the Aorta

Confirm Diagnosis

- Referral to Level 1 Centre
- Diagnosis confirmed via ECHO

Assessment

- Chest X-ray; Bloods,ECG; CMR; MVO2; Cardiac Catheter may be required to assess any other associated defects.
- Physical examination should include BP measured in upper and lower limbs
- If hypertension present then 24hr BP monitor should be arranged.

Intervention

- **Stenting** is the treatment of choice in adults in native CoA with appropriate anatomy
- For adults with recurring or residual CoA **angioplasty with or without stent implantation** can be effective.
- **Surgical treatment** in adults where necessary can be complicated and ascending to descending conduits may be preferable in cases of difficult anatomy. There is a risk of spinal cord injury with surgical repair of CoA.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods + ECHO within 6-8 weeks to assess for stent position and residual CoA.
- ECG to ensure sinus rhythm. If concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- All patients with CoA require regular follow up every 1-2 years. BP on upper and lower limbs should be assessed every visit.
- Imaging of Aorta with CMR (ot CT if CMR not feasible) should occur every 3-5 years to document the post op or post intervention anatomy and complications such as restenosis, stent fracture or migration, or aneurysm formation.
- Imaging may be required more frequently in known bicuspid aortic valve physiology or Shone syndrome.
- BP should be assessed at rest, ambulatory, or exercise test. If HTN, treatment should be start inmediatly.

11 Isolated Pulmonary Regurgitation after repair of Pulmonary Stenosis

Confirm Diagnosis

- Referral to Level 1 Centre
- TTE ECHO to assess PR severity and RV size and function

Assessment

- Chest X-ray; Bloods, ECG; CMR; MVO2

Intervention

- In symptomatic patients with moderate to severe PR and RV dilatation/dysfunction --> Pulmonary Valve Replacement (PVR) is recommended.
- In asymptomatic patients with moderate to severe PR with progressive RV dilation and/or RV dysfunction, PVR may be reasonable.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks to assess PVR, RVOT; RV function; and associated lesions.
- ECG to ensure sinus rhythm. I f concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- <u>Before intervention:</u>
- **Moderate or greater PR** and RV enlargement without symptoms and no evidence of progressive RV dilation and/or RV dysfunction and/or progressive decrease in exercise capacity - OPA with ACHD Team with ECG and ECHO every 12-24 months. (CMR and Exercise test as required).
- <u>After intervention:</u>
- **PVR and mild PR** - OPA with ACHD Team with ECG and ECHO every 24 months; (Exercise test as required)
- **PVR and significant PR or PS** - follow up with ECHO every 12months; (MRI/Exercise test as required)
- Endocarditis prophylaxis is recommended after PVR

12 Right Ventricular Outflow Tract Obstruction

Confirm Diagnosis

- Referral to Level 1 Centre
- RVOTO can occur at the subinfundibular; infundibular; valvular or supravalvular levels. Can be isolated or at multiple levels.
- RVOTO level, severity and RV funcion should be confirmed via TTE ECHO.

Assessment

- ECHO; Chest X-ray; Bloods, ECG; CMR; MVO2;
- Cardiac catheterisation may be required to assess severity of RVOTO
- V/Q scan may be required to assess lung perfusion abnormalities.
- Severity:
  - Mild RVOTO - gradient <36mmHg (Peak velocity <3m/s).
  - Mod RVOTO - gradient 36-64mmHg (Peak velocity 3-4m/s).
  - Severe RVOTO - gradient >64mmHg (Peak velocity >4m/s).

Intervention

- **Balloon valvuloplasty is the first choice in clasic domed** valvular PS and can even be considered in dysplastic type of valves.
- **Surgical valvulotomy or PVR** might be necesarry in patients with significant infundibular stenosis; hypolplastic pulmonary annulus; dysplastic pulmonary valves or other associated lesions such as severe PR or severe TR.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods + ECHO within 6-8 weeks to assess PVR, residual PS or PS, RVOTO; RV dysfunction;
- ECG to ensure sinus rhythm. If concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- **Prior to intervention:** If Moderate to Severe RVOTO, follow-up should be every 6 - 18 months depending on the severity of RVOTO, rate of progression and RV function
- **Post intervention follow-up:** every year for the first 2 years with ECHO and ECG.
- Patients with mild RVOTO without evidence of progression can be followed up every 3-5 years.
- CMR and MVO2 should be considered every 3-5 years depending on ECHO findings.
- Endocarditis prophylaxis is recommended after PVR

13 Branch and Peripheral Pulmonary Stenosis

Confirm Diagnosis

- Referral to Level 1 Centre
- TTE ECHO to confirm diagnosis and evaluate RV pressure and function

Assessment

- Bloods, ECG, MVO2 and Chest X Ray.
- Cardiac MRI / Cardiac CT scan to visualise anatomic and branch PA anatomy.
- Pulmonary Perfusion testing can be used to quantify relative pulmonary blood flow.

Intervention

- **Balloon angioplasty or stenting of peripheral PA** is effective in reducing pressure gradients and improving pulmonary blood flow. Indications for pulmonary angioplasty or stenting include symptoms attributed to decrease pulmonary blood flow, focal narrowing, abnormal differential perfusion, and/or elevated RV pressure.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods + ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological stage A - OPA with ACHD Team with ECG and ECHO every 24-36 months; Exercise Test every 36 months; CMR every 36-60 months.
- Physiological stage B - OPA with ACHD Team with ECG and ECHO every 24 months; Exercise Test every 24 months; CMR every 36-60 months.
- Physiological stage C: OPA with ACHD Team with ECG and ECHO every 6-12 months; Exercise Test every 24 months; CMR every 24-36 months.
- Physiological stage D - OPA with ACHD Team with ECG and ECHO every 3-6 months; Exercise Test every 12 months; CMR every 24-36 months.

14 Double-Chambered Right Ventricle

Confirm Diagnosis

- Referral to Level 1 Centre
- DCRV can be missed on TTE ECHO therefore CMR; CT; or Cardiac Catheter might be required to confirm diagnosis and establish the hemodynamic impact

Assessment

- Commonly associated with a VSD. Hypertrophied muscle bundles develop in the RV cavity, creating RVOT obstruction.
- Chest X-ray; Blood test, ECG; CMR; MVO2;
- If Echocardiogram and MRI differ - Cardiac Catheter may be required to assess for RV pressures and gradients.

Intervention

- **Surgical repair** for adults with DCRV and moderate or greater outflow obstruction is recommended in patients with otherwise unexplained symptoms of HF, cyanosis, or exercise limitation.
- **Surgical repair** for adults with DCRV with a severe gradient may be considered in asymtomatic patients.
- Surgical repair should contemplate closure of the VSD

Post Intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological stage A - OPA with ACHD Team; ECG and ECHO every 24-36 months. (Exercise Test as required).
- Physiological stage B - OPA with ACHD Team with ECG and ECHO every 24 months; Exercise Test every 24 months.
- Physiological stage C - OPA with ACHD Team every 6-12 months; ECG and ECHO every 12 months; Exercise Test every 24 months.
- Physiological stage D - OPA with ACHD Team every 3-6 months; ECG and ECHO every 12 months; Exercise Test every 12 months.

15 Right Ventricle to Pulmonary Artery Conduit

Confirm Diagnosis

- Referral to Level 1 Centre
- TTE ECHO to assess conduit anatomy and function and also valve function.

Assessment

- ECG, Chest Xray, Bloods, MVO2 and CMR.
- In patients with stented RV to PA conduits and worsening PS or PR, evaluation for conduit complications should be performed including fluoroscopy to evaluate for stent fracture and blood cultures to assess for IE.
- In adults with RV to PA conduit and arrhythmia, congestive HF, unexplained ventricular dysfunction or cyanosis cardiac catheterisation is reasonable to assess hemodynamics

Intervention

- Coronary artery compression testing with simultaneous coronary angiography and high pressure dilation in the conduit is indictated before RV to PA conduit stenting or transcatheter valve replacement.
- RV to PA conduit intervention is reasonable for adults with RV to PA conduit and moderate or greater PR or moderate or greater stenosis with reduced functional capacity or arrhythmia; or in asymptomatic patients with severe stenosis or severe regurgitation with reduced ejection fraction or RV dilation.

Post Intervention Follow-up

- Post procedure ECHO within 6-8 weeks. ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological stage A - OPA with ACHD Team with ECH and ECHO every 12-24 months; ECG and ECHO every 24 months; CMR every 36-60 months (Exercise Test as required).
- Physiological stage B - OPA with ACHD Team with ECG and ECHO every 12 months; CMR every 36-60 months (Exercise Test as required).
- Physiological stage C - OPA with ACHD Team every 6-12 months; ECG and ECHO every 12 months; CMR and Exercise Test every 12-24 months.
- Physiological stage D - OPA with ACHD Team every 3-6 months; ECG and ECHO every 12 months; CMR and Exercise Test every 12-24 months.

16 Congenital Mitral Stenosis

Confirm Diagnosis

- Referral to Level 1 Centre

Assessment

- Chest X-ray; ECG; Bloods, CMR; MVO2; TTE ECHO. Stress echo might be required.
- RH Catheter may be required to assess for any other associated defects.
- Complete assessment of other known associated congenital abnormalities, such as the components of Shone Syndrome

Intervention

- **Indication for valvce repair / replacement are as follows:**
- Significant mitrals tenosis (area <1.5 cm^2) in symptomatic patients; if asymptomatic, surgery should be considered if: history of systemic embolism, dense spontaneous contrast in the left atrium, new-onset atrial fibrillation, systolic PAP >50 mmHg at rest, need for major non-cardiac surgery or desire for pregnancy.
- Choices and techniques for valve repair or replacement are based on consideration of coexisting abnormalities.
- Congenital mitral stenosis is usually unsuitable for percutaneously baloon valvuloplasty.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered

Long-term Follow-up

- Recurrence and progression of other congenital abormalities is expected.
- Physiological Stage A - OPA with ACHD Team with ECG and ECHO every 24 months (Exercise test as required)
- Physiological Stage B - OPA with ACHD Team with ECG and ECHO every 24 months (Exercise test as required)
- Physiological Stage C - OPA with ACHD Team every 6-12 months with ECG and ECHO every 12 months. Exercise test every 24 months.
- Physiological Stage D - OPA with ACHD Team every 3-6 months with ECG and ECHO every 12 months. Exercise test every 12 months.

17 Cor Triatriatum

Confirm Diagnosis

- Referral to Level 1 centre
- Diagnosis should be confirmed confirmed via TTE ECHO.

Assessment

- Chest X-ray; Bloods, ECG; CMR; MVO2;
- Complete assessment should be made for other congenital abnormalities particularly ASD, VSD, and anomalous pulmonary venous connection (total or partial).
- Carefully rule out pulmonary venous stenosis - this has been demonstrated before and after surgery.
- Cardiac Catheter may be required during assessment.

Intervention

- Intervention should be considered in adults with cortriatriatum and symptoms attributable to the obstruction OR when there is a substantial gradient across the membrane (suggested gradient **is ≥8mmHg)**.
- Intervention can be considered with gradient <8mmHg if symptoms and/or arrhythmia are present.

Post intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered

Long-term Follow-up

- Follow-up is dependant on the presence of any other congenital abnormalities - if other abnormalities present then follow up should be according with the recommended follow-up for the congenital anomaly.
- If no other congenital abnormalities are present should be based on the residual gradient.
- Follow-up arranged for 12-36 months with ECG and ECG - according to consultant ACHD cardiologist.
- Exercise Test and CMR may be recommended during follow-up as per advice of consultant ACHD cardiologist.

18 (Partial) Anomalous Pulmonary Venous Drainage

Confirm Diagnosis

- Referral to Level 1 Centre
- Diagnosis of PAPVD confirmed on TTE ECHO

Assessment

- Chest X Ray, Bloods, MVO2 and ECG.
- Cardiac MRI or CT Angiography are recommended for evaluation of PAPVD.
- RH Cardiac Catheterisation can be used to further define haemodynamics.

Intervention

- **Surgical intervention for PAPVD** is recommended when impaired functional capacity and RV enlargement are present, there is a left to right shunt sufficient to cause physiological sequelae (i.e. Qp:Qs ≥1.5:1) with PAP <50% systemic pressure, and PVR <1/3 systemic resistance.
- If Sinus venosus ASD present, repair of PAPVD is recommended at time of Sinus venosus ASD closure.
- **Repair of Scimitar vein** is recommended if functional capacity is impaired, evidence of RV volume overload is present, left to right shunt sufficiently large to cause physiological sequelae (i.e. Qp:Qs ≥1.5:1), PAP <50% systemic pressure, and PVR <1/3 systemic resistance.
- In the absense of symptoms, it is reasonable to repair PAPVC and Scimitar vein if there are signs of RV volum overload and QP:Qs ≥1.5:1.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks to assess baffling of veins to LA or anastomosis sites, and also to check for SVC obstruction.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological Stage A - OPA with ACHD Team with ECG and ECHO every 36-60 months (Pulse Oximetry and Exercise test as required)
- Physiological Stage B - OPA with ACHD Team with ECG and ECHO every 24 months (Pulse Oximetry and Exercise test as required)
- Physiological Stage C - OPA with ACHD Team every 6-12 months with ECG and ECHO every 12 months. Pulse Oximetry at each visit and Exercise test every 12-24 months.
- Physiological Stage D - OPA with ACHD Team every 3-6 months with ECG and ECHO every 12 months. Pulse Oximetry at each visit and exercise test every 6-12 months.

19 Tetralogy of Fallot

• Referral to Level 1 Centre
•Residual lesions should be first assessed by Echo TTE

Diagnosis

• Chest X-ray; Blood test, ECG; MVO2.
• CMR should be used to quantify ventricular size and function, pulmonary valve function, pulmonary artery anatomy, and left heart abnormalities.
•Cardiac catheter with angiography is reasonable to assess hemodynamics when this cannot be assess non-invasively in the setting of arrhythmia, HF, unexplained ventricular dysfunction, suspected PHT or cyanosis.

Assessment

•In patients with risk factors, **consider refer to ARIA Clinic** for SCD Risk Assessment
•Programmed ventricular stimulation can be useful to risk-stratisfy adults with TOF and additional risk factors for sudden cardiac death.

•Consider Genetic Testing for DiGeorge Sd (specially in women with pregnancy desire)

Intervention

• **PVR (surgical or percutaneous)** is indicated in moderate or greater PR in:
  • For the reflief of symptoms
  • In aymptomatic patients for preservation of ventricular size and function with ventricular enlargement or dysfunction (RVEDV ≥160ml/m2, RVESV ≥80ml/m2, or RVEDV≥ 2x LVEDV); if RVOTO with RVSP ≥2/3 systemic; if the exercice capacity is declining in the MVO2.
  • Surgical PVR is reasonable if there are other lesions requiring surgery.
• PVR in addition to arrhythmia management may be considered for adults with repaired TOF and moderate or greater PR and ventricular tachycardia.

• Coronary artery compression testing is indicated before RV to PA conduit stenting or transcatheter valve replacement in repaired TOF

Intervention follow

• Post intervention CNS Review with Bloods and ECHO within 6-8 weeks.
• ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

•Physiological stage A - OPA with ACHD Team every 12-24 months; ECG and ECHO every 24 months; CMR every 36 months; Exercise test every 36-60 months (Pulse oximetry and Holter monitor as required).
•Physiological stage B - OPA with ACHD Team with ECG every 12 months; ECHO every 12-24 months; CMR every 24-36 months; Exercise Test every 24-60 months (Pulse Oximetry and Holter monitor as required).
•Physiological stage C - OPA with ACHD Team with Pulse Oximetry every 6-12 months; ECG and ECHO every 12 months; CMR, Exercise Test and Holter monitor every 12-24 months.
•Physiological stage D - OPA with ACHD Team with Pulse Oximetry every 3-6 months; ECHO every 6-12 months; ECG every 12 months; CMR, Exercise Tes, Holter monitor every 12-24 months.

20 Ebstein Anomaly

Confirm Diagnosis

- Referral to Level 1 Centre

Assessment

- In adults with Ebstein Anomaly ECHO and CMR are useful to determine anatomy, RV dimensions and systolic function.
- Request as baseline: Bloods, ECG, Chest X Ray and MVO2 (with Sat). Holter monitor if history of palpitations or syncopal episodes.
- Electrophysiology study with or without catheter ablation can be useful in the diagnostic evaluation of adults with Ebstein anomaly.

Intervention

- **Surgical repair or reoperation for adults with Ebstein anomaly and significant TR is recommended** when one or more of the following are present:
  - HF symptoms.
  - Objective evidence of worsening exercise capacity.
  - Progressive RV systolic dysfunction by ECHO or CMR.
- Surgical repair or reoperation for adults with Ebstein anomaly and sig TR **can be beneficial** in the presence of: progressive RV enlargement, systemic desaturation from right to left atrial shunt, paradoximal embolism, and/or atrial tachyarrhythmia.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods + ECHO within 6-8 weeks to assess TR and RV function;
- ECG to ensure sinus rhythm. If concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological stage A - OPA with ACHD Team with ECG and ECHO every 12-24 months; Pulse Oximetry every 24 months; Exercise Test every 36 months; CMR every 60 months (CXR and Holter Monitor as required).
- Physiological stage B - OPA with ACHD Team with ECG every 12 months; ECHO every 12-24 months; Pulse Oximetry every 12 months; Exercise Test every 24-36 months; CMR every 36 months (CXR and Holter Monitor as required).
- Physiological stage C - OPA with ACHD Team with Pulse Oximetry every 6-12 months; ECG and ECHO every 12 months; CXR every 12-24 months; Exercise Test every 24 months; CMR every 24-36 months; Holter Monitor every 24 months.
- Physiological stage D - OPA with ACHD Team with ECG and Pulse Oximetry every 3-6 months; ECHO every 12 months; Exercise Test every 12 months; CMR; Holter monitor and CXR every 12-24 months.

21 Truncus arteriosus

Confirm diagnosis

- Referral to level 1 Centre

Assessment

- ECG, CXR, echocardiogram, CMR, cardio-pulmonary exercise test, routine bloods
- Cardiac catheterization may be considered

Intervention

- Truncus arteriosus in the adult has almost invariably been repaired in childhood
- In the rare circumstances when an adult has unrepaired truncus arteriosus, Eisenmenger physiology is typical; Pulmonary hypertension may also be present in repaired patients
- The types of operative repairs may involve VSD closure, right ventricle–to-pulmonary artery conduit placement, reconstruction of the pulmonary arteries, and replacement of the truncal (neoaortic) valve
- Unifocalization of the pulmonary arteries may be necessary in very complex cases

Longterm follow-up

- Recommendations regarding assessment and management of truncus arteriosus are inferred in the recommendations for the specific components, including right ventricle–to-pulmonary artery conduit, VSD, aortic valve disease, aortopathies and Eisenmenger physiology
- Patients with Eisenmenger Physiology or Pulmonary Hypertension after repair should be referred to PH4 Clinic

22 Double Outlet Right Ventricle

Confirm diagnosis

- Referral to level 1 Centre

Assessment

- ECG, CXR, echocardiogram, CMR, cardio-pulmonary exercise test, routine bloods
- Cardiac catheterization may be considered
- Liver imaging (Ultrasonography with elastography, CMR, CT) and laboratory evaluation of liver function for fibrosis, cirrhosis, and/or hepatocellular carcinoma is required in patients with single-ventricle physiology and Fontan palliation

Intervention

- Double Outlet Right Ventricle is an anatomic descriptor that includes various abnormalities. Types of repairs are predicated on the underlying anatomy and may involve VSD closure with relief of pulmonary stenosis, right ventricle–to-pulmonary artery conduit, or Rastelli-type repair. In some cases, single-ventricle physiology may be present and Fontan palliation as a result performed

Longterm follow-up

- Guidelines for the management of a patient with double outlet right ventricle are inferred in the recommendations for the lesion with the most similar anatomy and physiology

23 Fontan

Confirm diagnosis

- Referral to level 1 Centre

Assessment

- ECG, CXR, echocardiogram, CMR, cardio-pulmonary exercise test, routine bloods
- Liver imaging (Ultrasonography with elastography, CMR, CT) and laboratory evaluation of liver function for fibrosis, cirrhosis, and/or hepatocellular carcinoma
- Cardiac catheterization may be required

Management

- New presentation of an atrial tachyarrhythmia in adults with Fontan palliation should be managed promptly and include prevention of thromboembolic events and consultation with an electrophysiologist with CHD expertise for a consideration of electrophysiology study and potential ablation
- Anticoagulation with a vitamin K antagonist is recommended for adults with Fontan palliation with known or suspected thrombus, thromboembolic events, prior atrial arrhythmia, atriopulmonary Fontan connection and no contraindications to anticoagulation
- In selected patients novel oral anticoagulant (NOAC) can be used if anticoagulation is recommended
- Antiplatelet therapy or anticoagulation with a vitamin K antagonist may be considered in adults after Fontan palliation without known or suspected thrombus, thromboembolic events, or prior arrhythmia and with total cavo-pulmonary connection (lateral tunnel or extracardiac conduit)
- Pulmonary vasoactive medications can be beneficial to improve exercise capacity in adults with Fontan repair
- Heart failure medications i.e. angiotensin converting enzyme inhibitors, angiotensin receptor blockers and mineralocorticoid receptor antagonists may be used with a potential benefit in adults after Fontan palliation with cardiac dysfunction

Intervention

- Fontan revision surgery, including arrhythmia surgery as indicated, may be considered for adults with atriopulmonary Fontan connections with recurrent atrial tachyarrhythmias refractory to pharmacological therapy and catheter ablation who have preserved systolic ventricular function, no valvular disease, low pulmonary artery pressures (mPAP<15mmHg) and severe atrial dilation
- Reoperation or intervention for structural/ anatomic abnormalities in a Fontan palliated patient with symptoms or with failure of the Fontan circulation may be considered
- Heart transplant may be considered in Fontan patients with cardiac dysfunction, Fontan pathway dysfunction, lymphatic dysfunction or extracardiac dysfunction as specified in heart transplant referral pathway

Longterm follow-up

- Physiological Stage A - OPA with ACHD Team with ECG and ECHO every 12 months, CMR/CT every 36months, Cardiopulmonary eercise Test every 36 months
- Physiological Stage B - OPA with ACHD Team with ECG and ECHO every 12 months, CMR/CT every 24months, Exercise test every 24 months.
- Physiological Stage C - OPA with ACHD Team with ECG every 6months, ECHO every 12 months. CMR /CT every 24 months, Cardiopulmonary exercise test every 12 months.
- Physiological Stage D - OPA with ACHD Team withh ECG every 3-6 months , ECHO every 12 months, CMR/CT every 24 months, ECardiopulmonary eercise test every 12 months.
- Imaging of the liver (ultrasonography with elastography, CMR, CT) and laboratory evaluation of liver function for fibrosis, cirrhosis, and/or hepatocellular carcinoma every 1-2 years in adults after Fontan palliation
- Bochemical and hematological testing every 12 months especially for liver and renal function
- Cardiac catheterization should be performed in adults before initial Fontan surgery or revision of a prior Fontan connection to assess suitability of preintervention hemodynamics for Fontan physiology or revision of a prior Fontan connection
- New onset or worsening atrial tachyarrhythmias in adults with single ventricle after Fontan palliation should prompt a search for potential hemodynamic abnormalities, which may necessitate imaging and/or cardiac catheterization
- Cardiac catheterization can be useful to evaluate a symptomatic adult after Fontan palliation when noninvasive testing is insufficient to guide therapy
- New presentation of an atrial tachyarrhythmia in adults with Fontan palliation should prompt consultation with an electrophysiologist with CHD expertise

24 Transposition of the Great Arteries – Atrial Switch

Confirm Diagnosis

- Referral to Level 1 Centre

Assessment

- Chest X-ray; Blood test, ECG; MVO2
- TTE Echocardiogram and CMR to assess specially RV size and function, tricuspid valve function, subpulmonic obstruction, obstruction of systemic or pulmonary venous return, and baffle leaks.
- Holter monitor should be considered, as there is a progressive loss of sinus rhythm, particularly in the context of beta-blocker treatment.

Intervention

- Atrial tachycardias should be addressed immediately, as they can be a potential cause for SCD in these patients.
- Efforts to maintain sinus rhythm or atrial pacing (and not simply rate control) should be the initial strategy
- Assessment of SCD risk and ICD implantation should be considered in collaboration with the EP team

Post Intervention Follow-up

- Post intervention CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological stage A - OPA with ACHD Team with ECG and pulse oximetry every 12 months; ECHO every 12-24 months; Holter monitor every 24 months; CMR every 24-36 months; Exercise Test every 36 months;
- Physiological stage B - OPA with ACHD Team with ECG and pulse oximetry every 12 months; ECHO every 12-24 months; Holter monitor 24 months; CMR every 24 months; Exercise Test every 36 months;
- Physiological stage C - OPA with ACHD Team with ECG and pulse oximetry every 6-12 months; ECHO every 12 months; Holter monitor every 12 months; CMR every 12-24 months; Exercise Test every 24 months.
- Physiological stage D - OPA with ACHD Team with ECG and pulse oximetry every 3-6 months; ECHO every 12 months; Holter monitor every 12 months; CMR every 12-24 months; Exercise Test every 12 months.

- Patients with RV dysfunction and HF symptoms should be referred for Heart Transplant Assessment.

25 Transposition of the Great Arteries – Arterial Switch

Confirm Diagnosis

- Referral to Level 1 Centre

Assessment

- Chest X-ray; Blood test, TTE Echocardiogram, ECG; CMR; MVO2
- Baseline assessment of the coronary arteries with CTCA/coronary catheterization should be performed
- Physiological exams of myocardial perfusion (like stress echo, nuclear test) should be performed in symptomatic patients

Intervention

- **Coronary revascularization** should be based on the current Revasc Guidelines and the Interventional Team should be envolved
- Complications of the arterial switch, **such as RVOT obstruction** (supravalvular PS), might require catheter or surgical intervention if symptoms of HF or decreased exercise capacity are present.
- Dilatation of neoaorta or signficant aortic regurgitation might need to be adressed surgically.

Post Intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological stage A - OPA with ACHD Team with ECG and ECHO every 12-24 months; Exercise Test and MRI every 36 -60 months months;
- Physiological stage B - OPA with ACHD Team with ECG every 12 months; ECHO every 12-24 months; CMR every 24-36 months; Exercise Test every 36-60 months;
- Physiological stage C - OPA with ACHD Team ECG every 6-12 months; ECG and ECHO every 12 months; CMR every 12-24 months; Exercise Test every 24-36months.
- Physiological stage D - OPA with ACHD Team with ECG every 3-6 months; ECHO every 12 months; CMR and Exercise Test every 12 months;

26 Congenitally Corrected Transposition of the Great Arteries

Confirm Diagnosis

- Referral to Level 1 Centre

Assessment

- Chest X-ray; Blood test, ECG; CMR; MVO2
- TTE Echocardiogram, specially oriented on the RV size and function and tricuspid valve anatomy and function.
- Holter monitor should be considered, as the prevalence of spontaneous complete heart block increases with age.

Intervention

- **Triscupid valve replacement** (or repair in selected patients) is indicated in symptomatic patients with severe TR and preserved (or only mildly impaired) RV impairment.
- In asymptomatic patients, tricuspid valve replacement can be considered in patients with severe TR, RV dilatation and mild RV dysfunction.
- In patients with LV-PA conduit dysfunction, conduit replacement might be considered (but unloading the LV might negatively impact the RV function).

Post Intervention Follow-up

- Post surgery CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Physiological stage A - OPA with ACHD Team with ECG and ECHO every 12 months; Holter monitor every 12-60 months; Exercise Test and CMR every 36 -60 months months; Pulse oximetry as needed.
- Physiological stage B - OPA with ACHD Team with ECG and ECHO every 12 months; Holter monitor every 12-60 months; Exercise Test and CMR every 36 -60 months months; Pulse oximetry as needed.
- Physiological stage C - OPA with ACHD Team with ECG and pulse oximetry every 6-12 months; ECHO every 12 months; Holter monitor every 12-36 months; CMR and Exercise Test every 12-24 months.
- Physiological stage D - OPA with ACHD Team with ECG and pulse oximetry every 3-6 months; ECHO every 12 months; Holter monitor every 12 months; CMR and Exercise Test every 12 months.

- Patients with RV dysfunction and HF symptoms should be referred for Heart Transplant Assessment.

27 Anomalous Aortic Origin of Coronary Artery

Confirm Diagnosis

- Referral to Level 1 Centre

Assessment

- **CTCA** should be performed to help identify high risk anatomic features like fish-mouth-shaped or slit-like orifice, or intramural course.
- **Physiological study** should be performed with a Stress Echo
- Other basic investigations: Chest X-ray; Blood test, TTE Echo, ECG; CMR; MVO2
- Coronary catheterization (with IVUS) might be required.

Intervention

- **Surgical repair** for adults should be considered in the presence of ischemia symptoms or if the stress test is positive for ischemia or arrythymias
- **Surgery or continued observation** may be reasonable for asymptomatic patients **without** ischemia or anatomic or physiological evaluation suggesting potential for compromise

Post Intervention Follow-up

- Post procedure CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long-term Follow-up

- Every 12 months (with functional test) if not repaired
- Every 24-36 months (with functional test if appropriate) if repaired.

28 PH and Eisenmenger Syndrome

Confirm Diagnosis

- Referral to Level 1 Centre
- Patients should be assessed by the ACHD and PH Team

Assessment

- Chest X-ray; Blood test, ECG; MVO2 or 6 min walking test
- TTE Echocardiogram and CMR to assess specially RV size and function.
- RHC to confirm pulmonary vascular hemodynamics and to exclude other potential contributors to right-to-left shunting or pulmonary hypertension.

Intervention

- Therapeutic treatment is indicated in patients with FC II, III or IV WHO

Long-term Follow-up

- Patients with Pulmonary Hypertension or Eisenmenger Syndrom should be referred to PH4 Clinic.
- Physiological stage C - OPA with ACHD Team with ECG and pulse oximetry every 6-12 months; ECHO every 12 months; CMR as needed; Exercise Test (CPET or 6 min WT) every 6-12 months.
- Physiological stage D - OPA with ACHD Team with ECG and pulse oximetry every 3-6 months; ECHO every 12 months; CMR as needed; Exercise Test (CPET or 6 min WT) every 6-12 months.
- Patients with HF symptoms should be referred for Heart and Lung Transplant Assessment.
- Cyanotic patients --> Endocarditis Prophylaxis for invasive dental procedures is recommended

29 Reference

Stoute *et al.* 2018 AHA / ACC Guideline for the Management of Adults with Congenital Heart Disease. Circulation. 2019; 139.