



A lifetime of specialist care

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

Adapted by L Shaughnessy, May 2019. Circulated and checked by the adult congenital heart disease consultant group May 2019. Ratified by the CHD ODN Board and CHD ODN Pathways & Protocols Sub-Committee June 2019.



A lifetime of specialist care

1 Contents

1	Contents.....	2
2	Physiological States.....	3
3	Atrial Septal Defect.....	4
4	Ventricular Septal Defect.....	5
5	Atrioventricular Septal Defect.....	6
6	Patent Ductus Arteriosus.....	7
7	Congenital Valvular Aortic Stenosis.....	8
8	Subaortic Stenosis.....	9
9	Supravalvular Aortic Stenosis.....	10
10	Coarctation of the Aorta.....	11
11	Isolated Pulmonary Regurgitation after repair of Pulmonary Stenosis.....	12
12	Right Ventricular Outflow Tract Obstruction.....	13
13	Branch and Peripheral Pulmonary Stenosis.....	14
14	Double-Chambered Right Ventricle.....	15
15	Right Ventricle to Pulmonary Artery Conduit.....	16
16	Congenital Mitral Stenosis.....	17
17	Cor Triatriatum.....	18
18	(Partial) Anomalous Pulmonary Venous Drainage.....	19
19	Tetralogy of Fallot.....	20
20	Ebstein Anomaly.....	21
21	Truncus arteriosus.....	22
22	Double Outlet Right Ventricle.....	23
23	Fontan.....	24
24	Transposition of the Great Arteries – Atrial Switch.....	26
25	Transposition of the Great Arteries – Arterial Switch.....	27
26	Congenitally Corrected Transposition of the Great Arteries.....	28
27	Anomalous Aortic Origin of Coronary Artery.....	29
28	PH and Eisenmenger Syndrome.....	30
29	Reference.....	31



A lifetime of specialist care

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

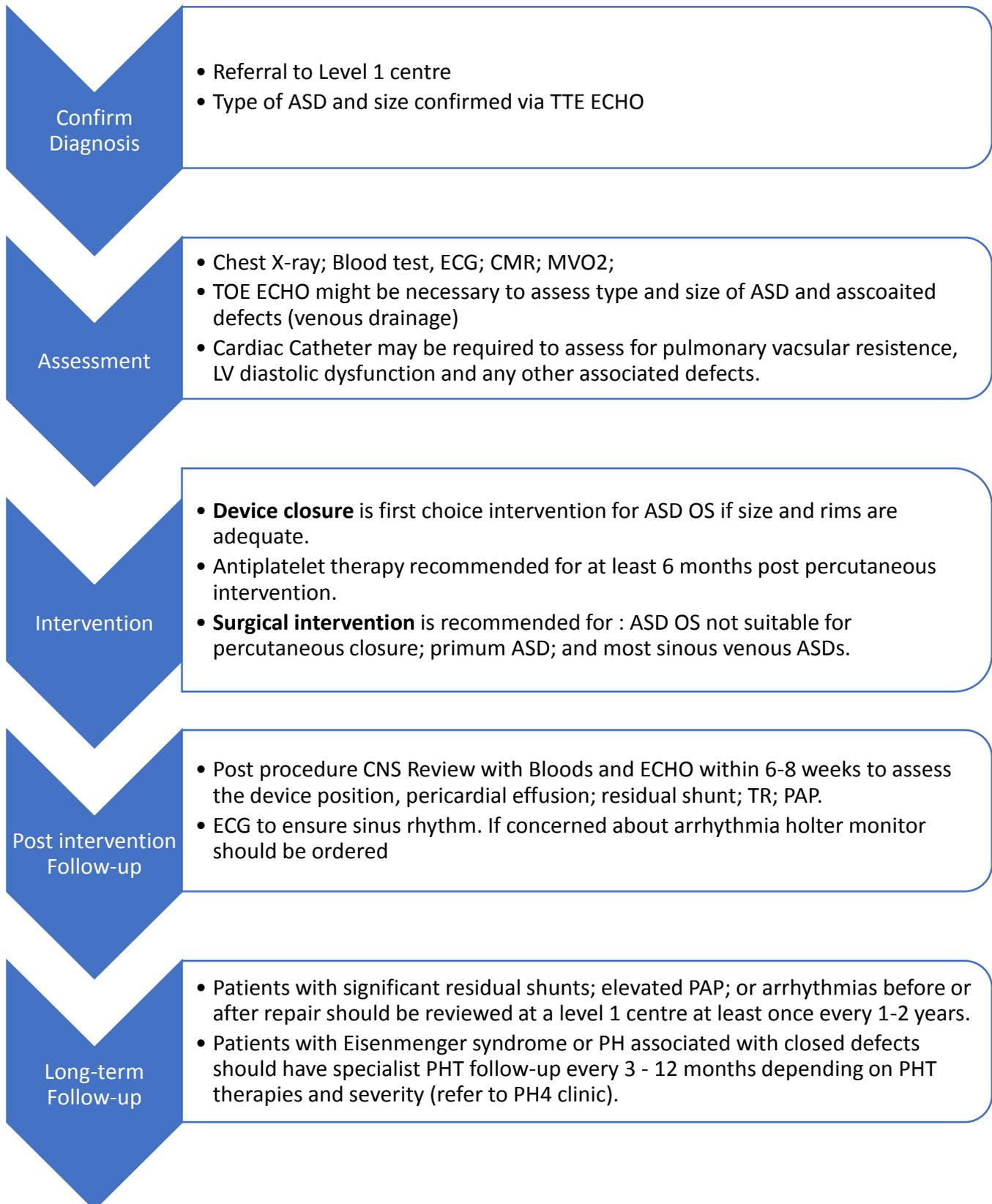
D

- 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



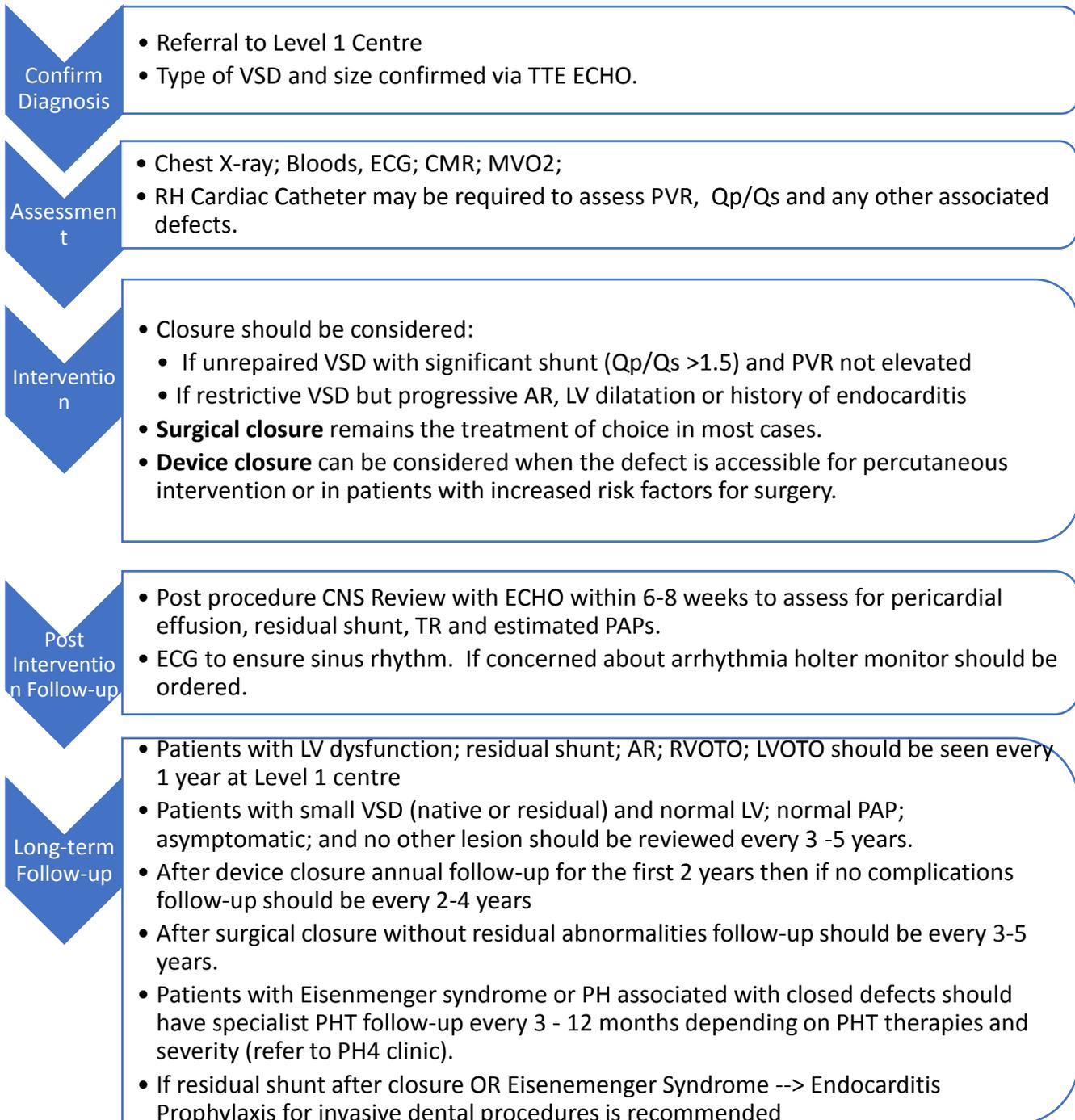
A lifetime of specialist care

3 Atrial Septal Defect





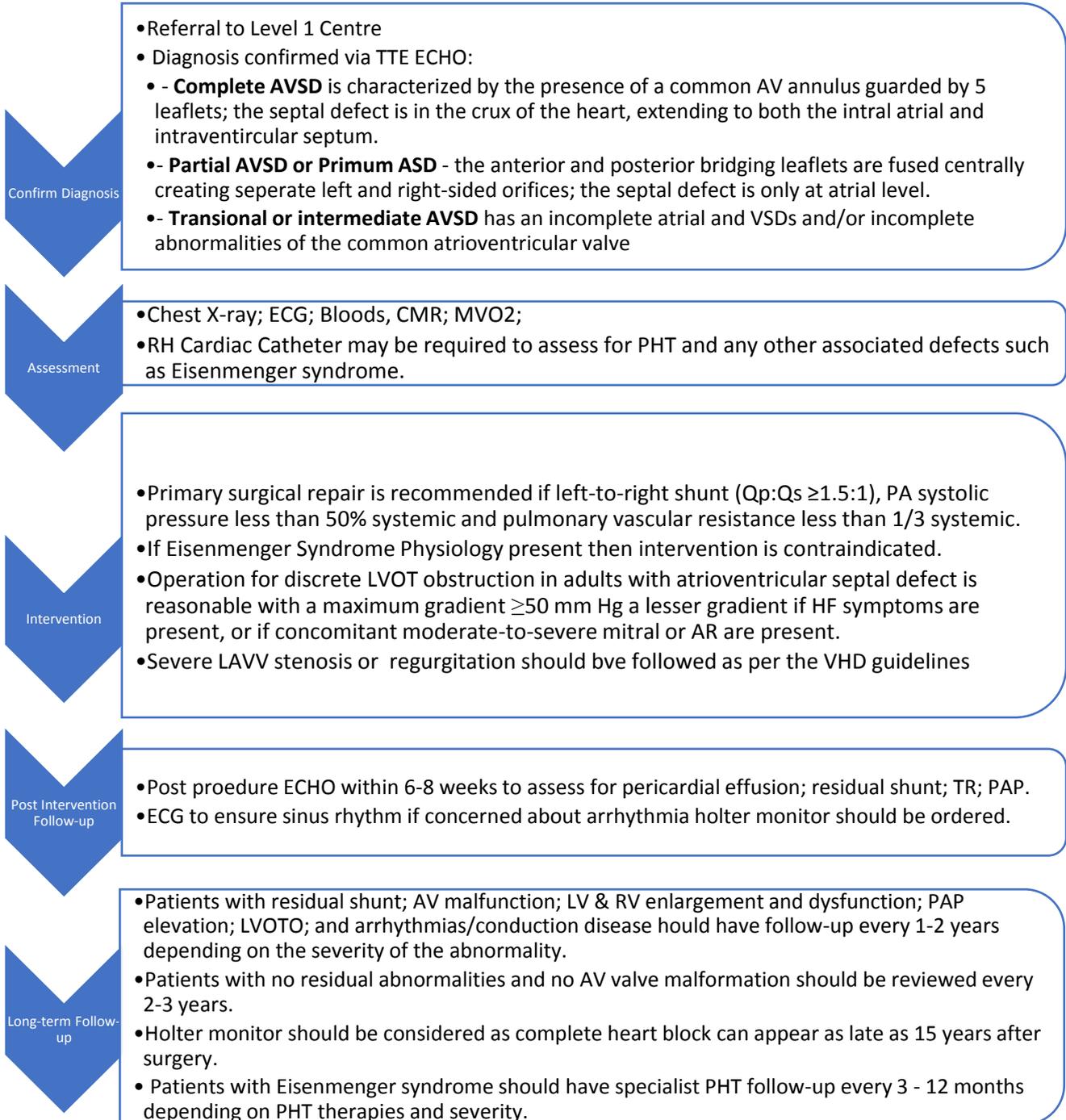
4 Ventricular Septal Defect





A lifetime of specialist care

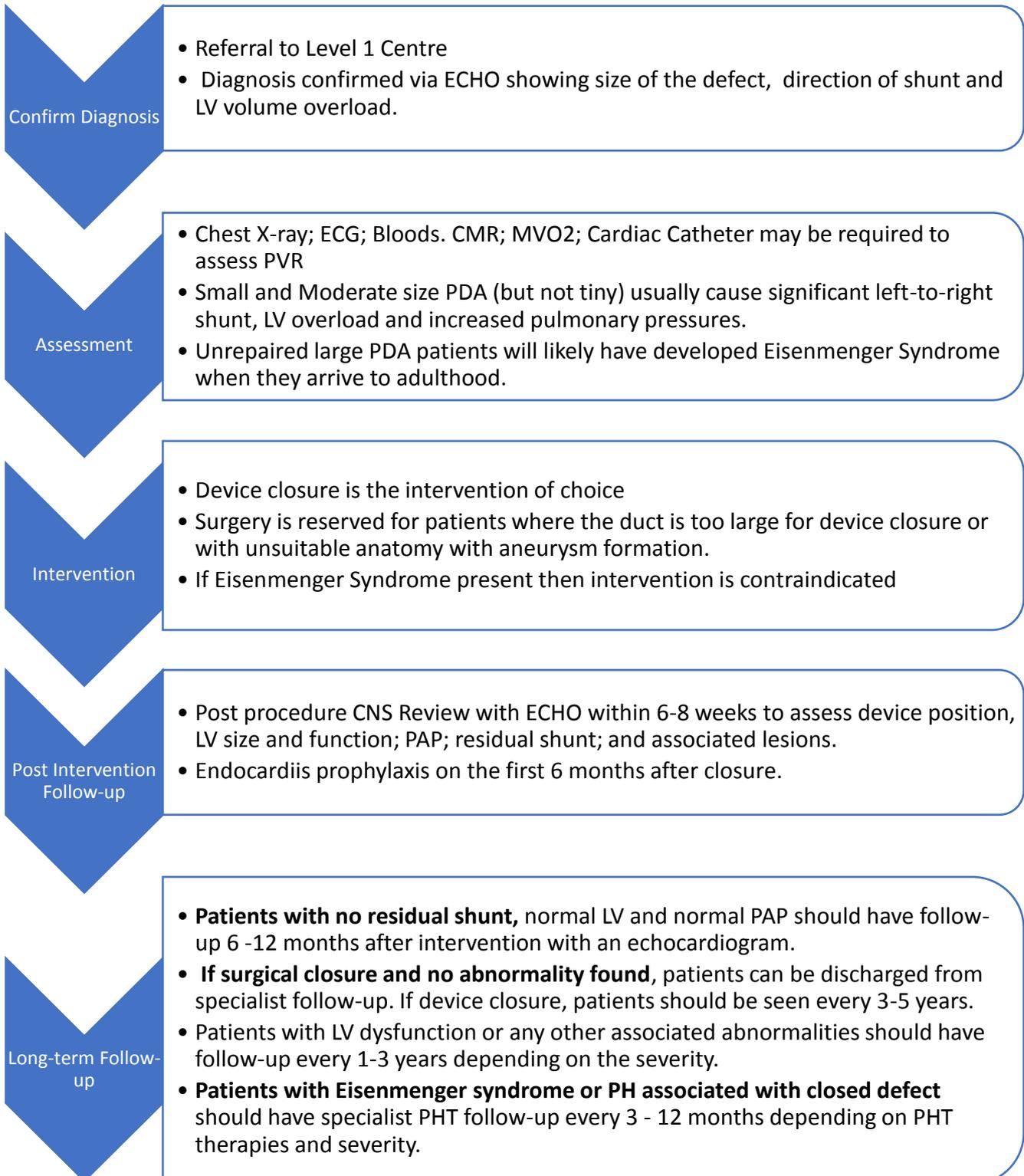
5 Atrioventricular Septal Defect





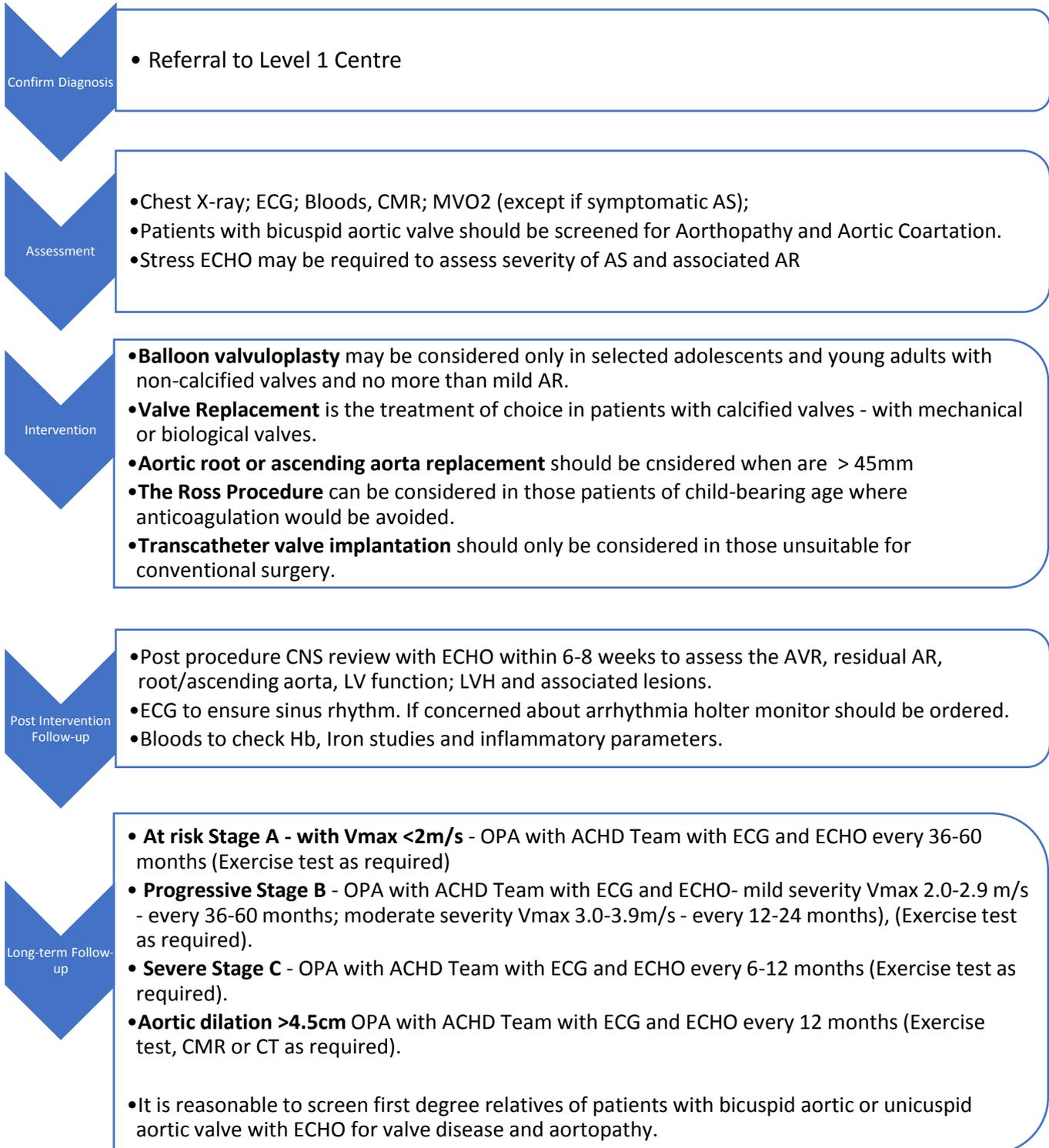
A lifetime of specialist care

6 Patent Ductus Arteriosus



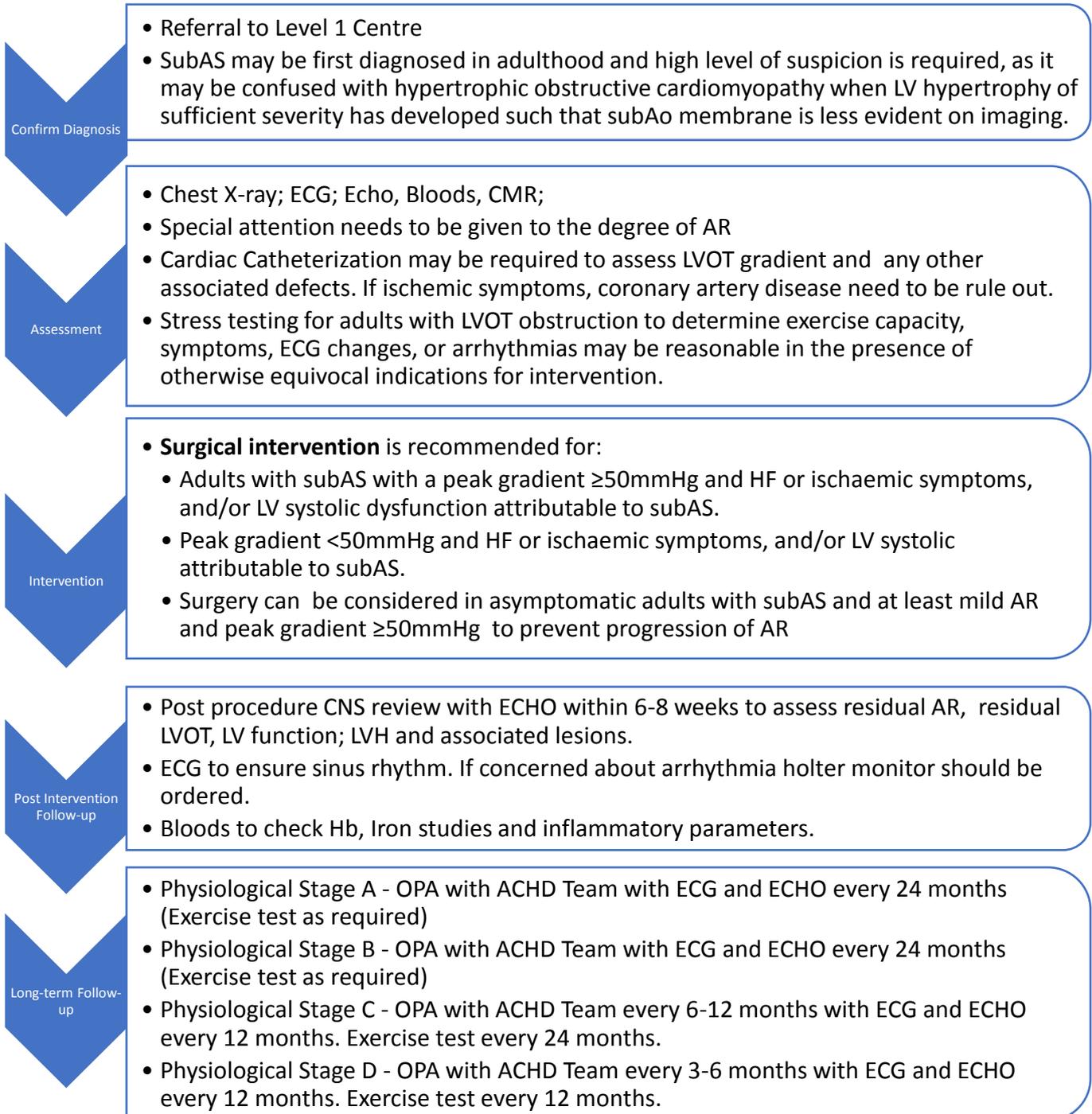


7 Congenital Valvular Aortic Stenosis





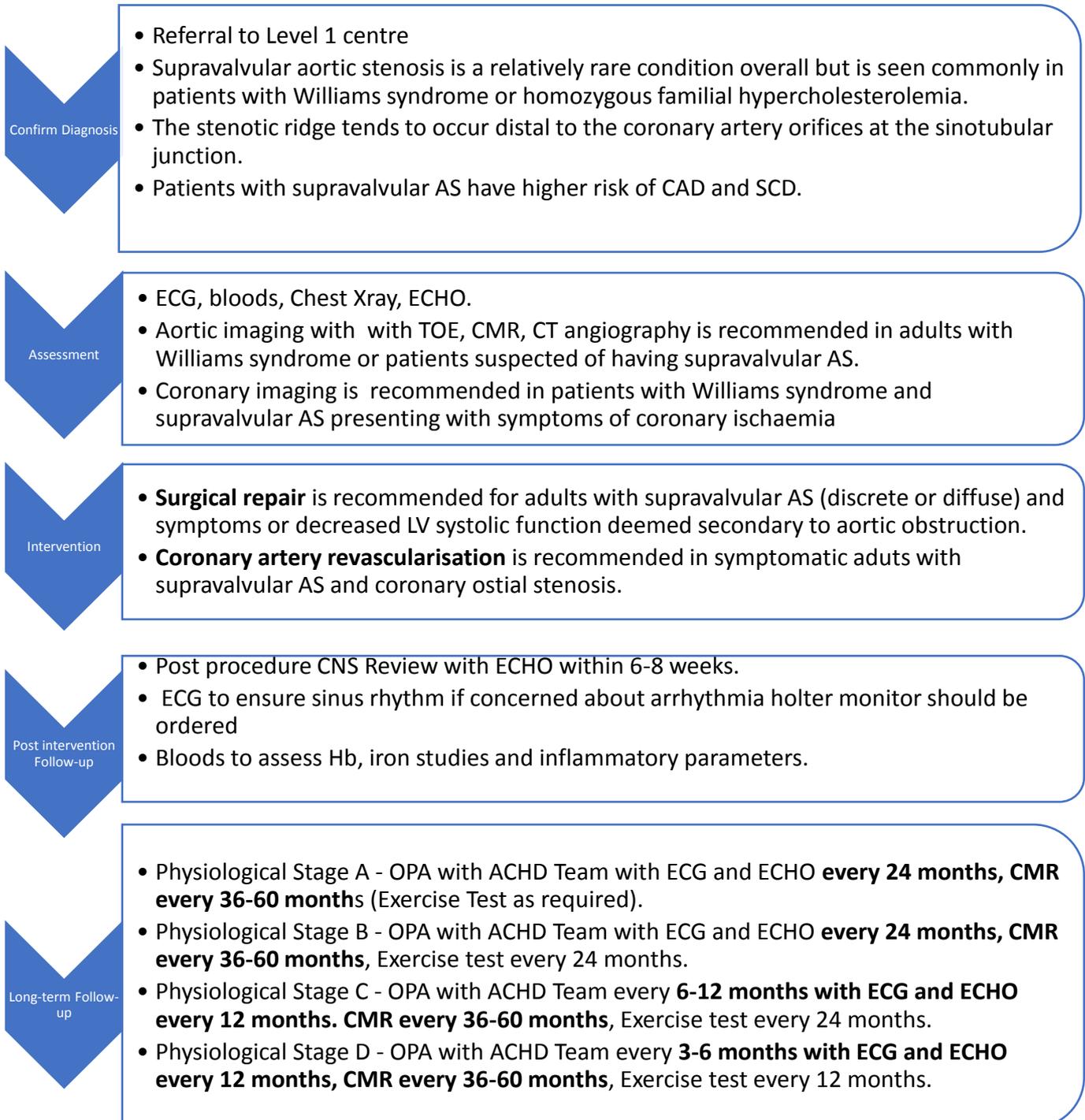
8 Subaortic Stenosis





A lifetime of specialist care

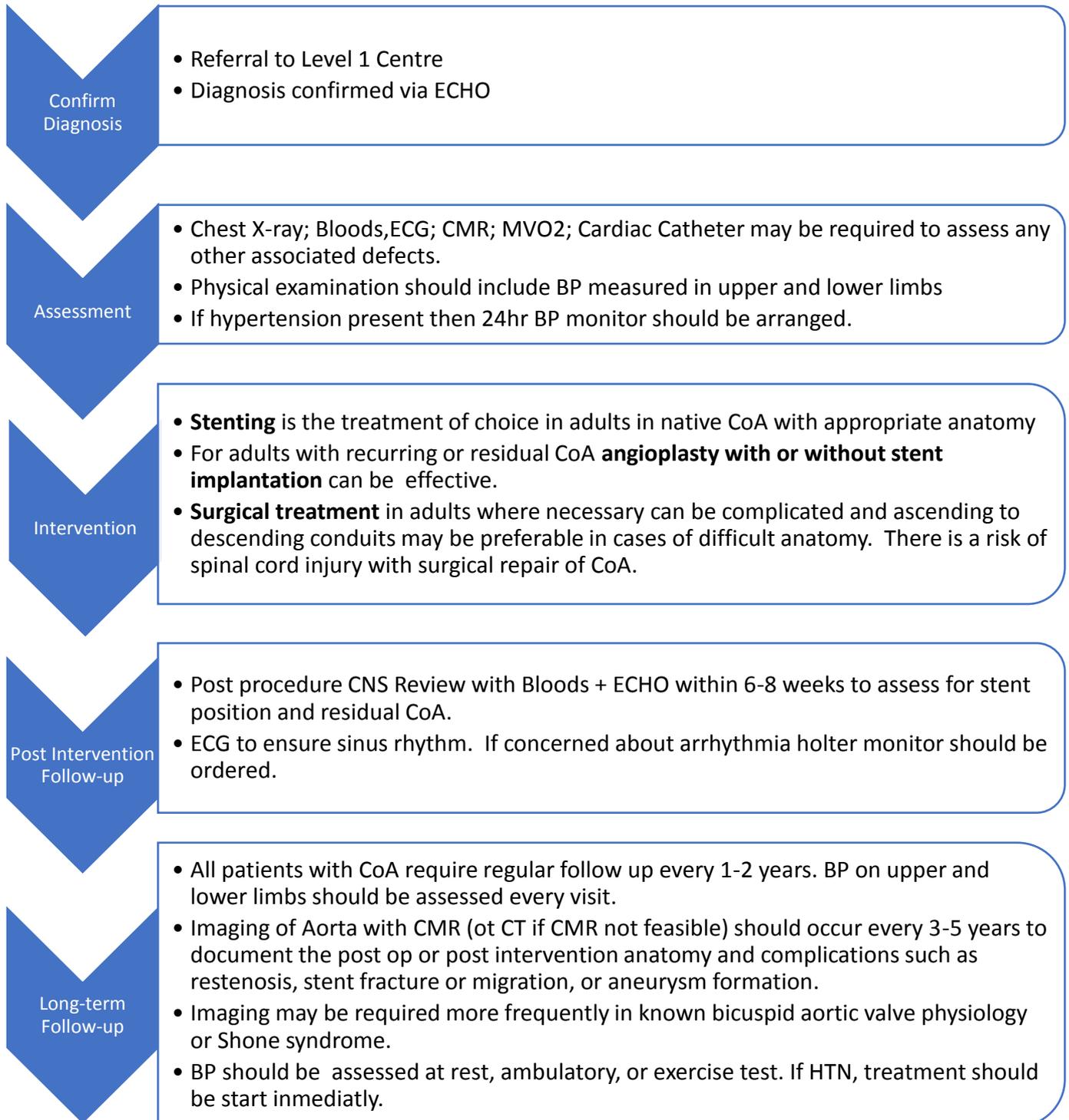
9 Supravalvular Aortic Stenosis





A lifetime of specialist care

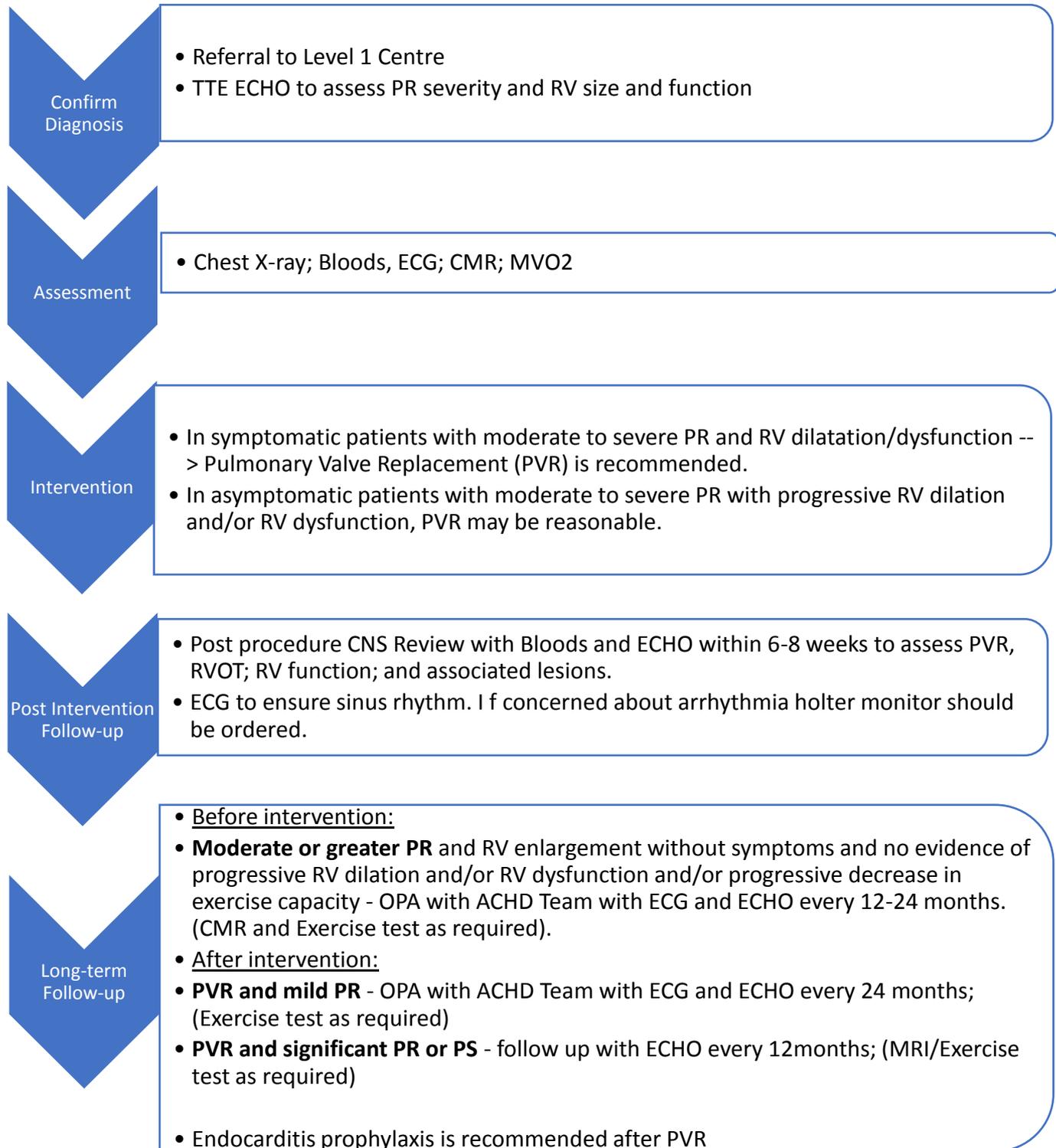
10 Coarctation of the Aorta





A lifetime of specialist care

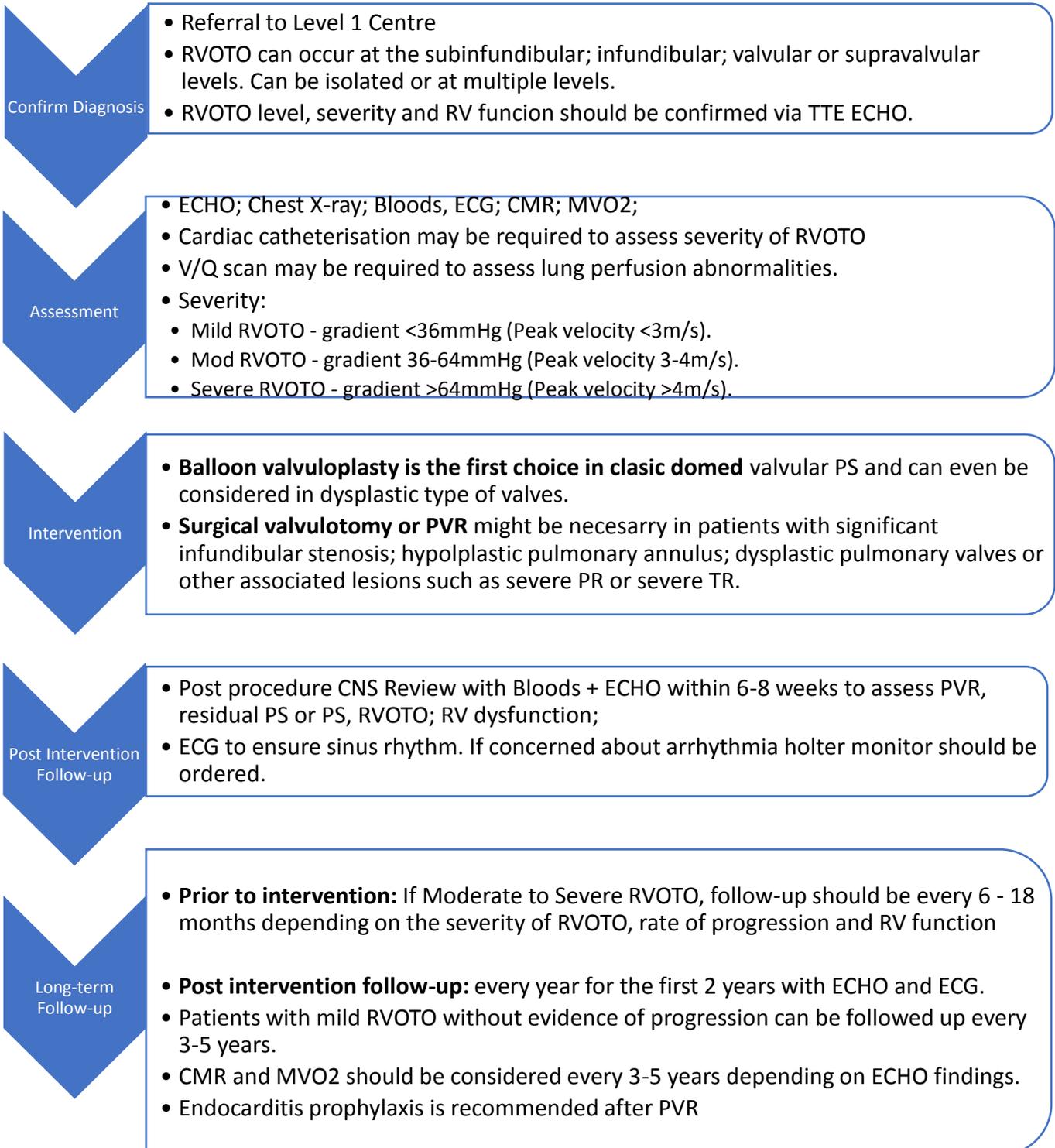
11 Isolated Pulmonary Regurgitation after repair of Pulmonary Stenosis





A lifetime of specialist care

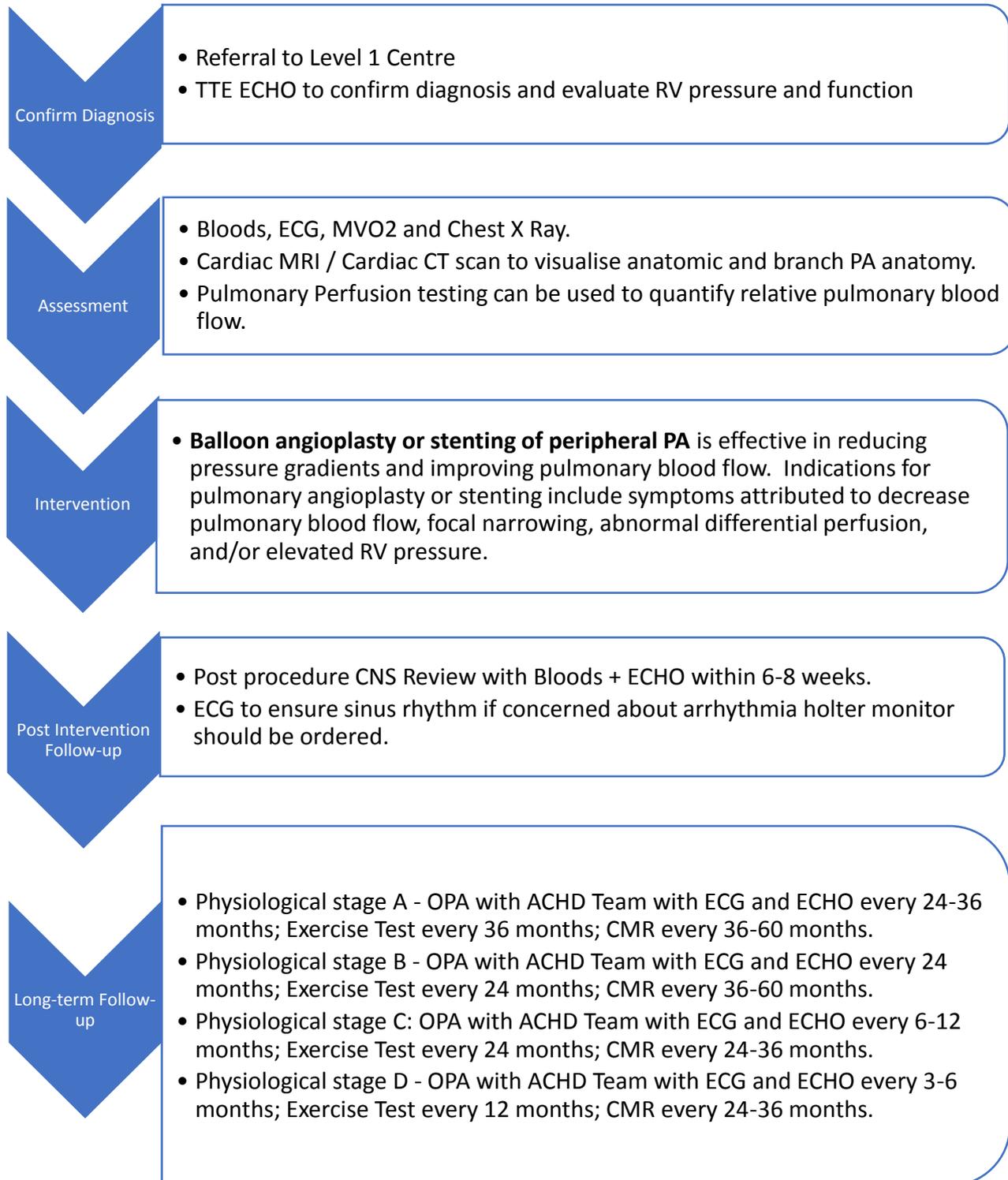
12 Right Ventricular Outflow Tract Obstruction





A lifetime of specialist care

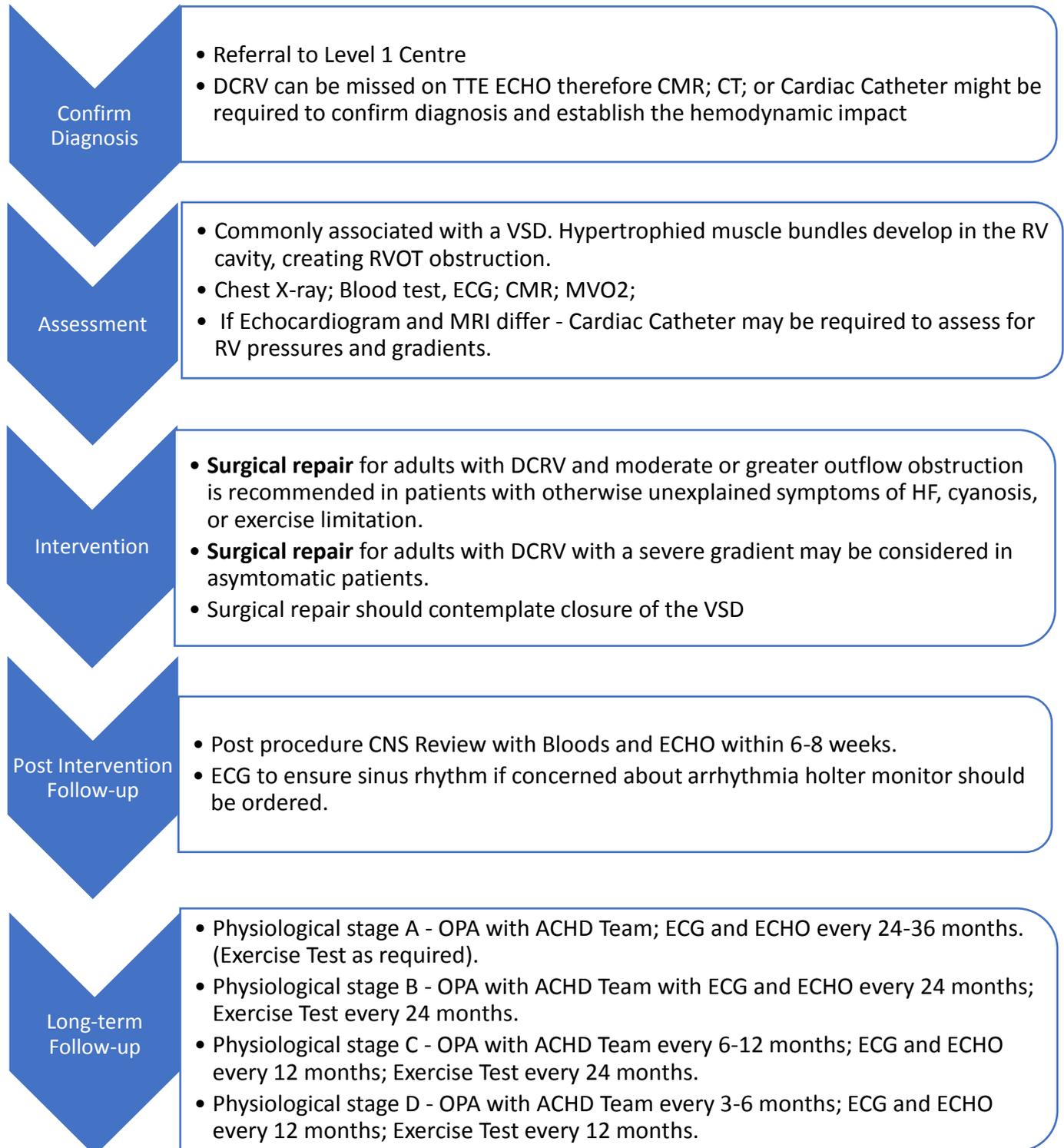
13 Branch and Peripheral Pulmonary Stenosis





A lifetime of specialist care

14 Double-Chambered Right Ventricle





A lifetime of specialist care

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 indicated 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

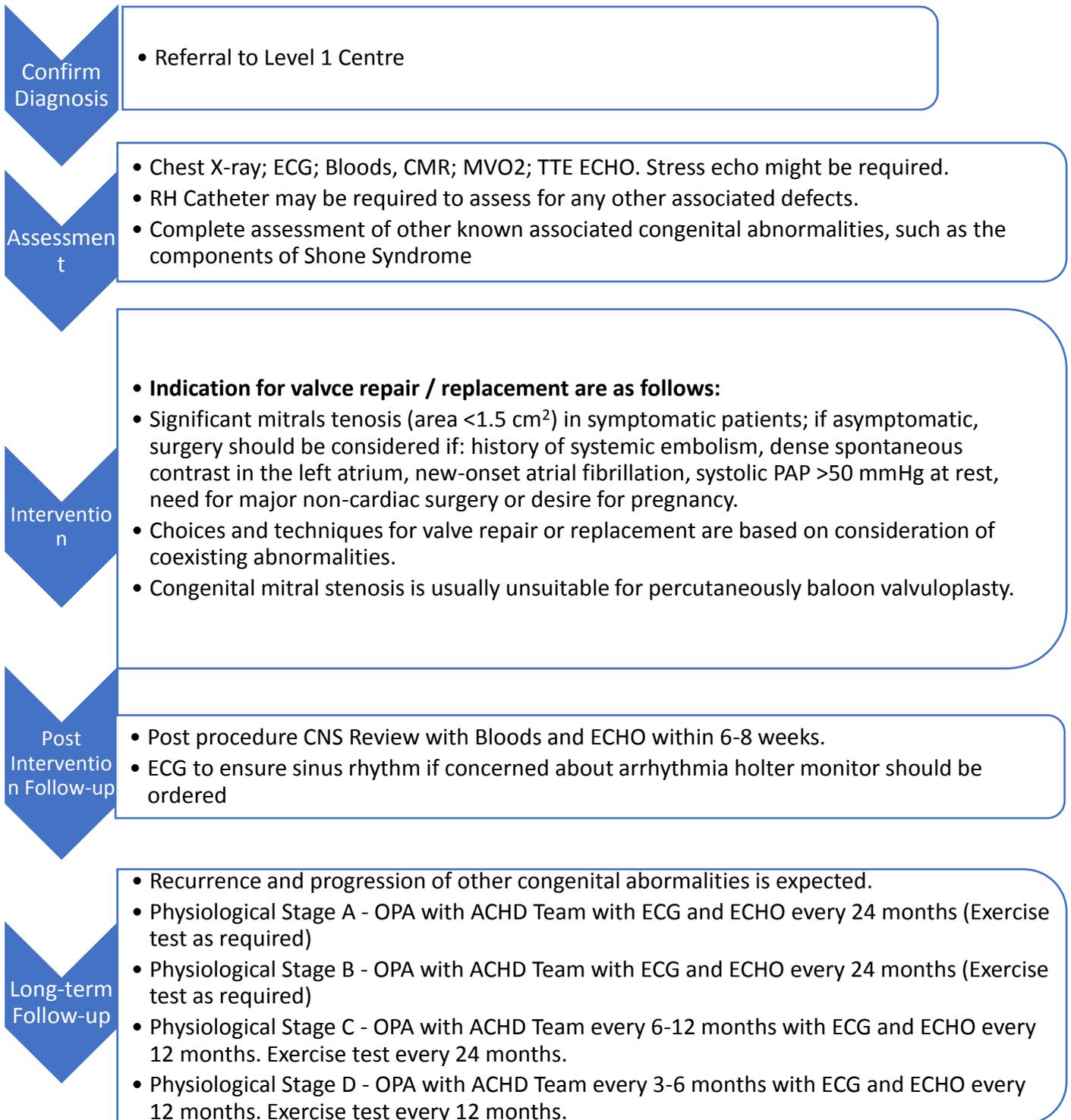
- Post procedure ECHO within 6-8 weeks. ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

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.

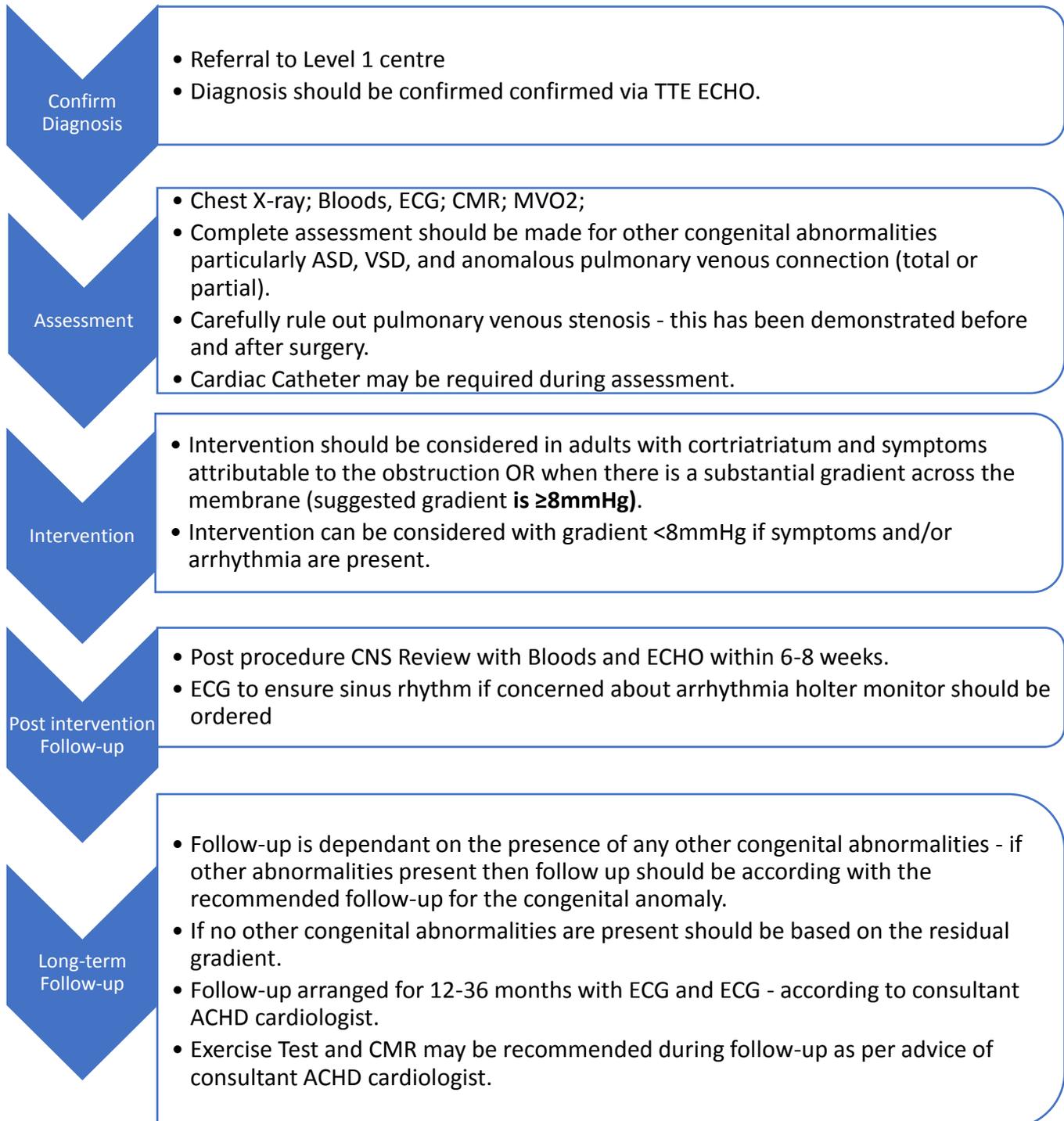


16 Congenital Mitral Stenosis



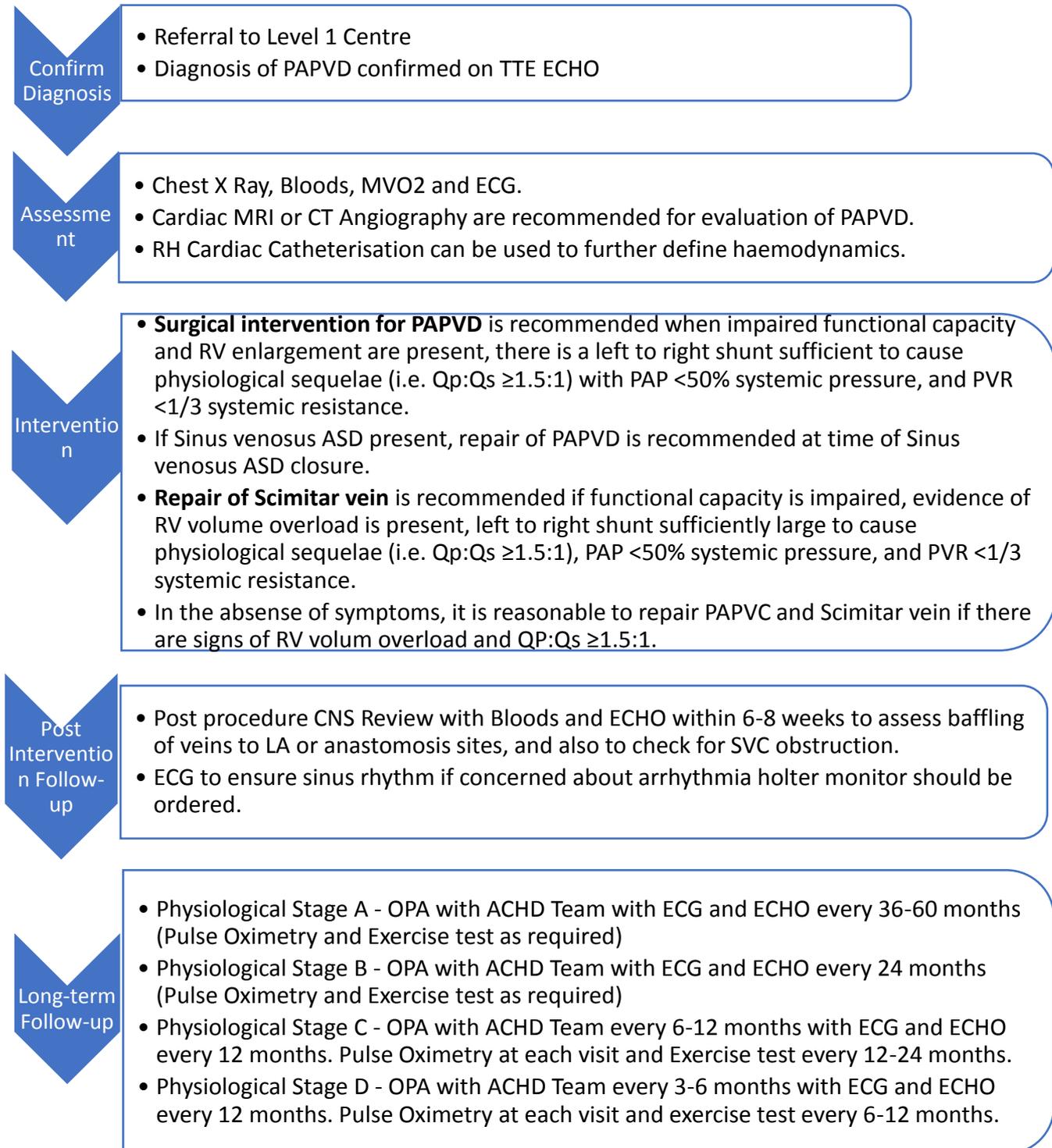


17 Cor Triatriatum





18 (Partial) Anomalous Pulmonary Venous Drainage





A lifetime of specialist care

19 Tetralogy of Fallot

Confirm
Diagnosis

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

Assessment

- 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 assessed non-invasively in the setting of arrhythmia, HF, unexplained ventricular dysfunction, suspected PHT or cyanosis.
- In patients with risk factors, **consider refer to ARIA Clinic** for SCD Risk Assessment
- Programmed ventricular stimulation can be useful to risk-stratify 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 relief of symptoms
 - In asymptomatic patients for preservation of ventricular size and function with ventricular enlargement or dysfunction (RVEDV ≥ 160 ml/m², RVESV ≥ 80 ml/m², or RVEDV $\geq 2 \times$ LVEDV); if RVOTO with RVSP $\geq 2/3$ systemic; if the exercise 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

Post
Intervention
Follow-up

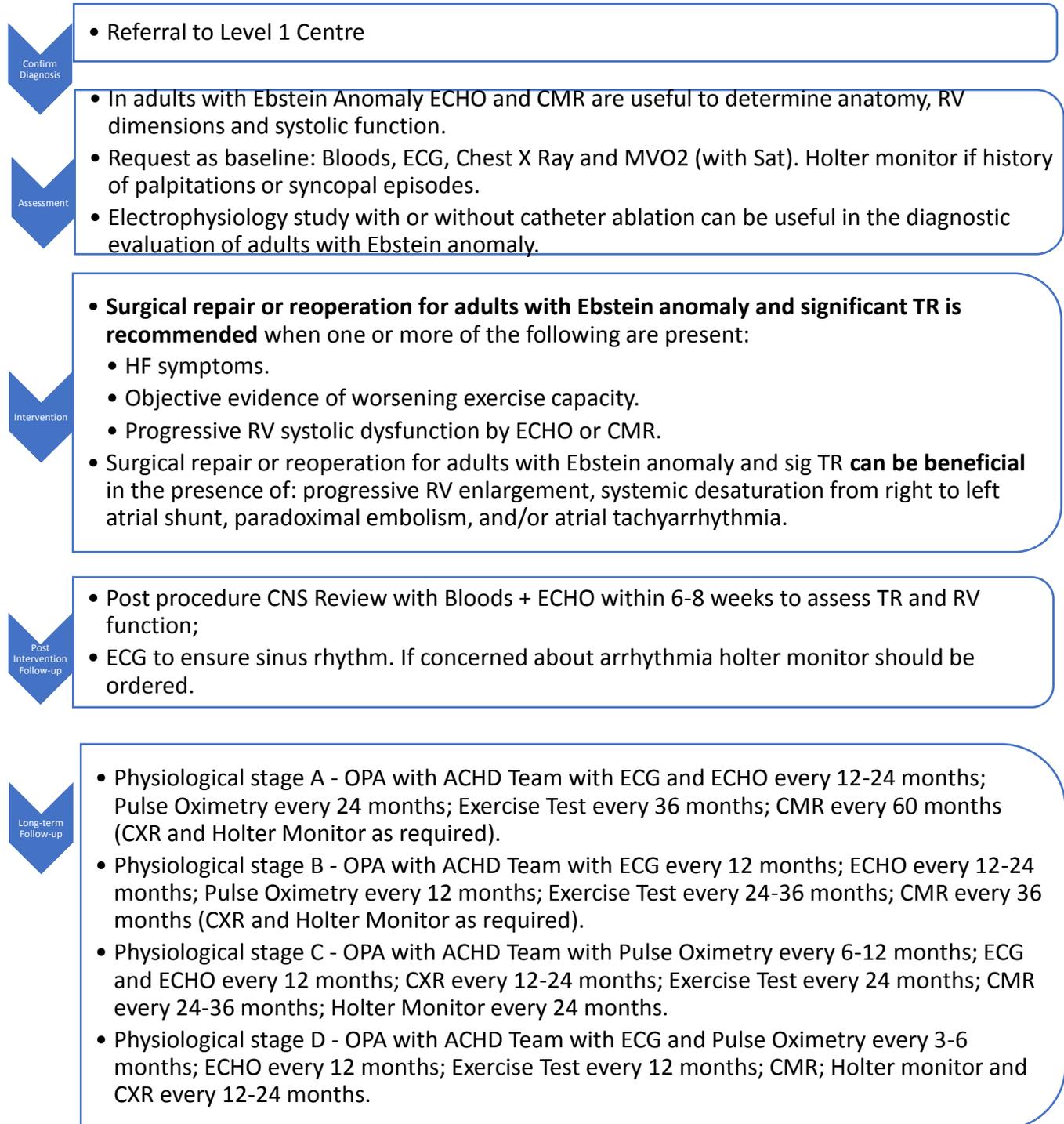
- Post intervention CNS Review with Bloods and ECHO within 6-8 weeks.
- ECG to ensure sinus rhythm if concerned about arrhythmia holter monitor should be ordered.

Long
term
Follow-up

- Physiological stage A - OPA with ACHD Team 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 Test, Holter monitor every 12-24 months.



20 Ebstein Anomaly





A lifetime of specialist care

21 Truncus arteriosus





A lifetime of specialist care

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



A lifetime of specialist care

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



A lifetime of specialist care

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 < 15 mmHg) 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

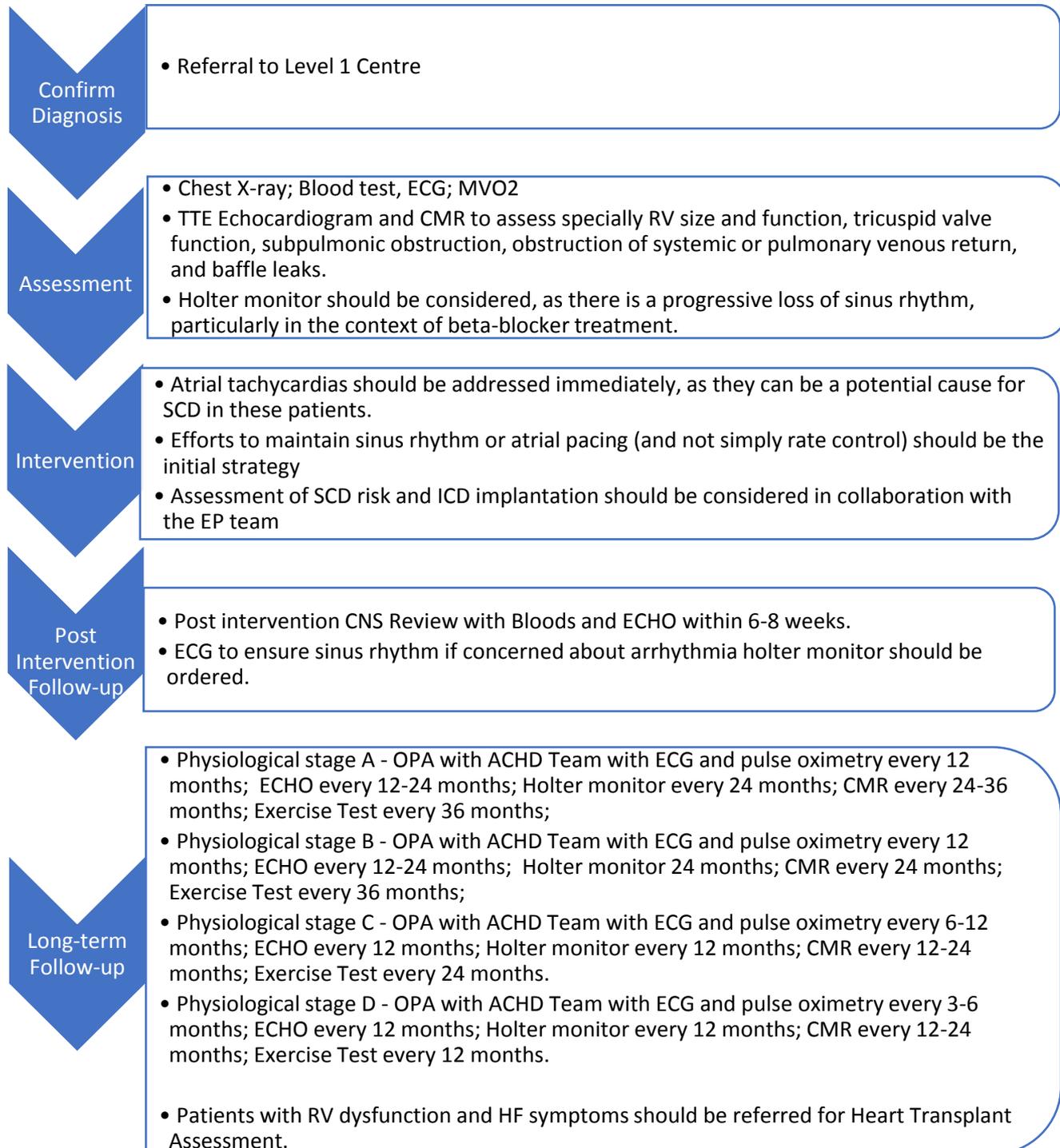
Longterm follow-up

- Physiological Stage A - OPA with ACHD Team with ECG and ECHO every 12 months, CMR/CT every 36 months, Cardiopulmonary exercise Test every 36 months
- Physiological Stage B - OPA with ACHD Team with ECG and ECHO every 12 months, CMR/CT every 24 months, Exercise test every 24 months.
- Physiological Stage C - OPA with ACHD Team with ECG every 6 months, ECHO every 12 months. CMR /CT every 24 months, Cardiopulmonary exercise test every 12 months.
- Physiological Stage D - OPA with ACHD Team with ECG every 3-6 months, ECHO every 12 months, CMR/CT every 24 months, ECardiopulmonary exercise 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
- Biochemical 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



A lifetime of specialist care

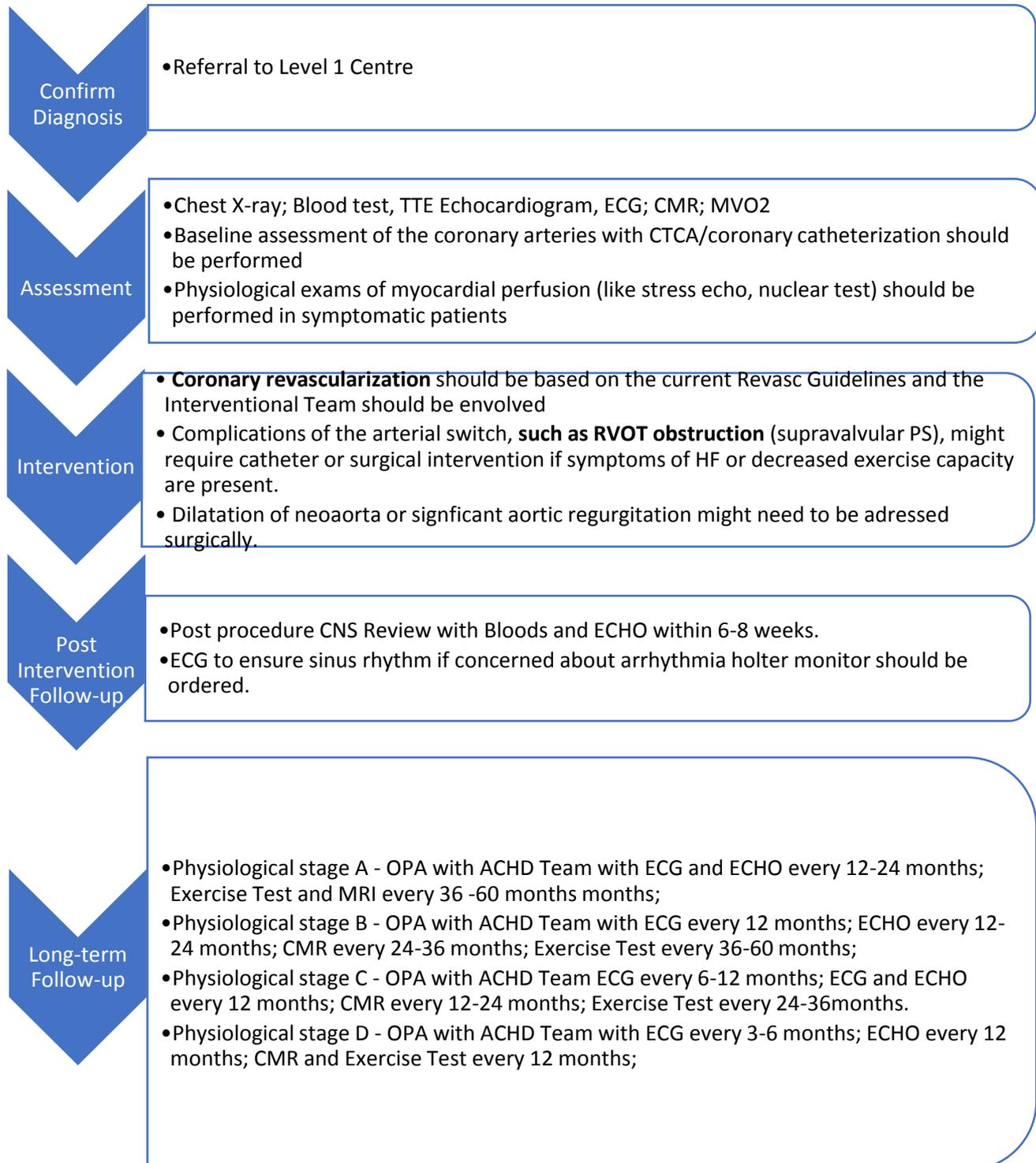
24 Transposition of the Great Arteries – Atrial Switch





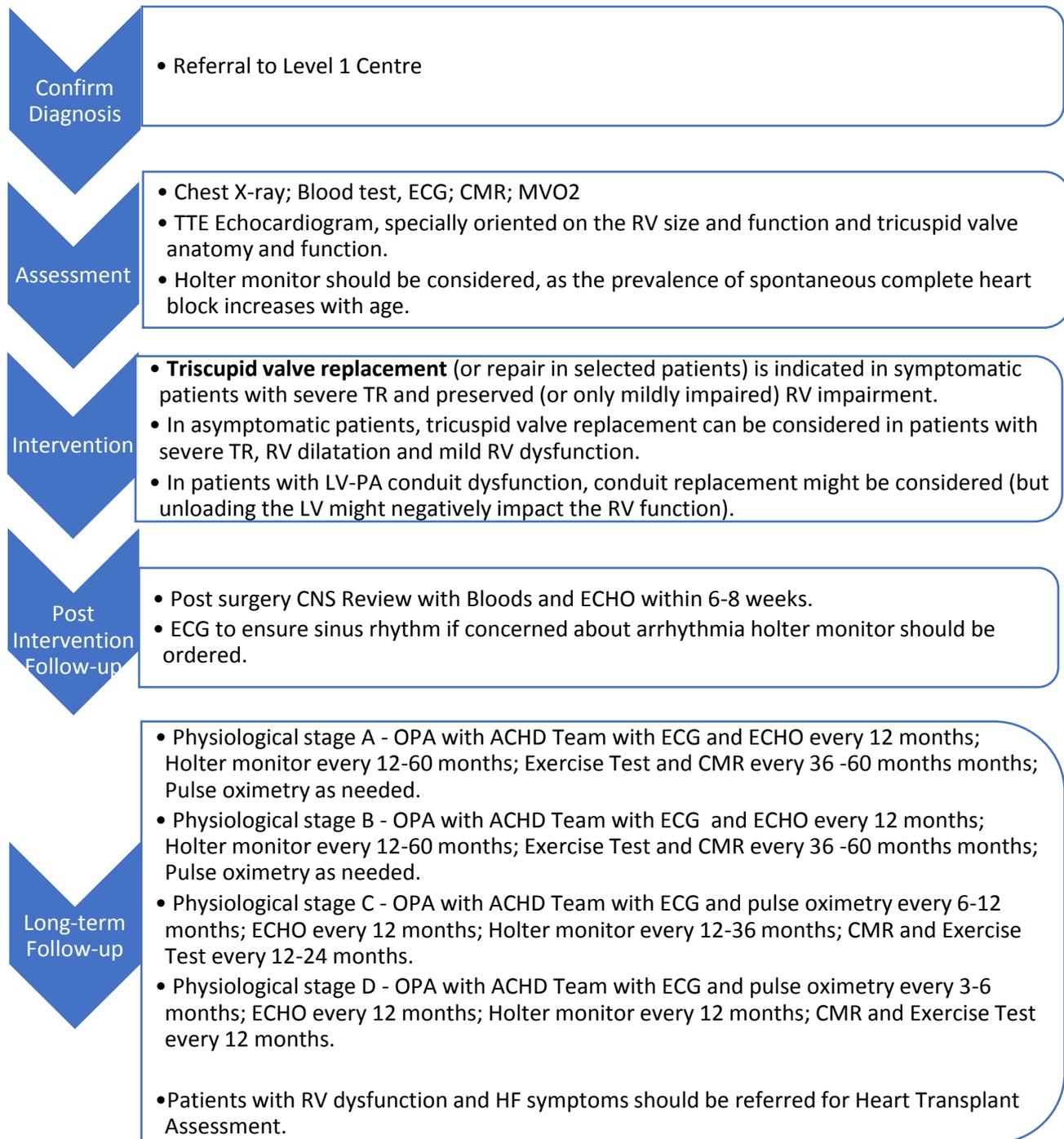
A lifetime of specialist care

25 Transposition of the Great Arteries – Arterial Switch





26 Congenitally Corrected Transposition of the Great Arteries





A lifetime of specialist care

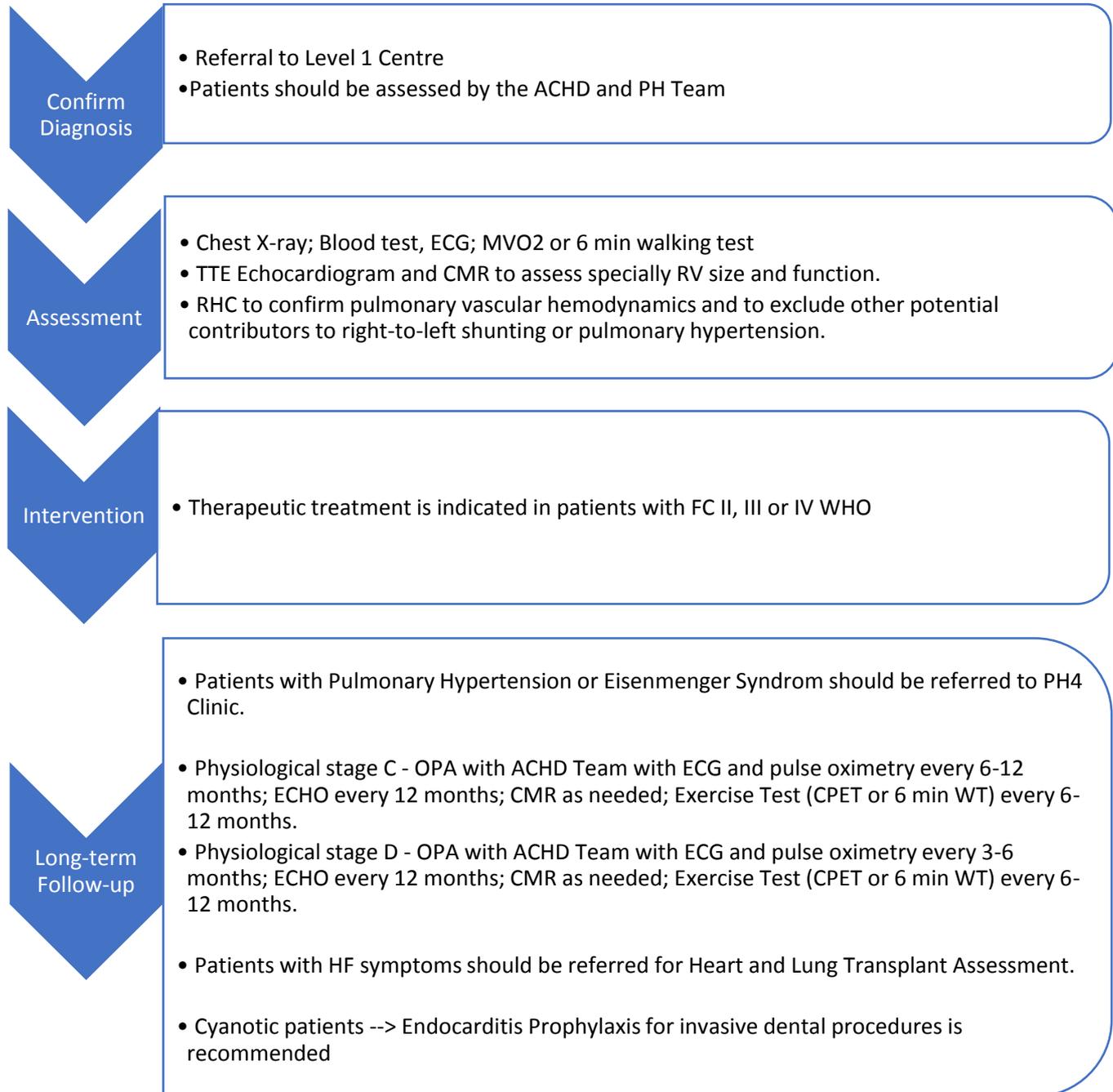
27 Anomalous Aortic Origin of Coronary Artery





A lifetime of specialist care

28 PH and Eisenmenger Syndrome





A lifetime of specialist care

29 Reference

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