A lifetime of specialist care

Royal Brompton & Harefield NHS
NHS Foundation Trust

Clinical Recommendations for Paediatric Cardiology

Royal Brompton Hospital Congenital Heart Disease Network

These are the agreed recommendations from the Royal Brompton Hospital Paediatric Cardiology 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

Written by AG Magee June 2009. 1st Revision by N Naqvi, Michael Rigby & Rodney Franklin February 2011, 2nd Revision N Naqvi – circulated and checked by cardiology consultant group May 2019. Ratified by the CHD ODN Board and CHD ODN Pathways & Protocols Sub-Committee June 2019.

1 Contents

2 Introduction

The following are management recommendations for children with congenital heart disease. They are not proscriptive but meant as guidelines, recognizing that individualization is necessary in many cases. They do not include every type of cardiac disorder but address the more common lesions and other common cardiac non-congenital conditions.

3 Nomenclature

3.1 Basic Investigations

This refers to a clinical examination, plus or minus ECG and Chest X-ray (all tests at the cardiologist's final discretion for example: consideration might be given to not performing routine Chest X-Rays at post-op visit if pre-discharge X-Ray satisfactory). For all cyanotic patients this would also include oxygen saturation at time of clinic assessment, plus haemoglobin and haematocrit at discretion of cardiologist.

3.2 Other Investigations

CT and Magnetic resonance imaging

For more complex aortic arch anomalies and for older children with complex anatomy, CT or CMR may be considered if inadequate imaging by echo. CMR is also becoming the investigation of choice in the assessment of RV function for example after repair Tetralogy of Fallot while CT adds to our understanding of coronary anatomy.

Exercise testing

MVO2 exercise testing can be a helpful objective measure of cardiovascular efficiency, and is particularly useful in serial follow-up of patients and for timing of interventions such as pulmonary valve replacement following Tetralogy of Fallot repair. Standard exercise testing or Thallium scanning is preferred in the investigation of ischaemia.

Karyotyping

This should be performed in all conotruncal lesions and those with a right aortic arch looking for 22q11 deletion, as well as lymphocyte subsets before surgery in these patients (if awaiting karyotype results) to decide if irradiated blood products are required (see 22q11 guidelines on Trust intranet). and indeed, there should be a low threshold for other complex lesions or where there are lesions specific to particular syndromes, such as subaortic stenosis and Williams Syndrome, etc.

4 Timing of First Follow-Up Visit

In general terms the first follow-up visit after surgery should be at 2-3 weeks after discharge mainly to check for pericardial effusion. If clinics are full, the effusion check should be as a day case with a formal outpatient appointment at a later date. After a catheter intervention the first follow-up is usually at 3 months although this may be sooner at the Cardiologist's discretion.

5 Anomalous Left Coronary Artery from the Pulmonary Artery

Preoperative studies – All require echo prior to surgical re-implantation. CT or cardiac catheter may be performed if the diagnosis by echocardiography is unclear however caution must be exercised if ventricular function is poor.

Postoperative studies

	Basic	Echo	Myocardial Perfusion
pre discharge	*	*	
2-3 weeks	*	*	
3 months	*	*	
12 months	*	*	
every 1-2 yrs	*	*	individualize

An exercise test is likely to be useful in older children or adolescents, as well as stress echo or stress CMR if any symptoms or concerns about coronary ischaemia. Myocardial perfusion scan (nuclear medicine or CMR) may be indicated if there are concerns about function on echo.

6 Anomalous Pulmonary Venous Drainage (total).

Preoperative studies

Echo only with typical findings. + Angiography, CMR, or CT angiography may be necessary if unable to see all 4 pulmonary veins by echo and there is a possibility of mixed drainage.

Postoperative studies

	Basic	Echo	Holter
pre discharge	*	*	
2-3 weeks	*	*	
3 months	*	*	
6 months	*	*	
1 year	*	*	*
3-5 years	*	individualise	*preschool, and at ages 10 & 13 & at transfer to ACHD care (1)

+ = Further investigations individualized based on echo result

Angiography, CMR, or CT angiography may be necessary if there is any evidence of pulmonary venous obstruction.

At age 10 consider as well as ECG & echo to perform exercise testing, repeat Holter, MRI and screening cholesterol.(1)

7 Aortic Valve Stenosis

Preoperative studies

All require echo prior to intervention.

Yearly follow-up for those ≥ 2 years, 6 monthly < 2 years.

If > 8 years of age then exercise ECG may be considered if estimated *peak to peak* gradient is ≥ 40 mmHg but less than 50 mmHg, unless there is ischemia on resting ECG.

The spectral recordings resulting from Doppler interrogations of aortic stenosis reflect the highest gradients within the jet. This is frequently referred to as 'peak instantaneous gradient'. Doppler estimates of the aortic peak gradient calculated using the simplified Bernoulli's formula generally overestimate severity when compared with catheterization peak-to-peak gradients. The mean gradient on echo is similar to the 'peak to peak' gradient and should be routinely recorded..

Indications and type of intervention

Neonatal Period (< 1 month age): Balloon dilatation to be considered unless otherwise contraindicated by valve morphology. JCC to decide on strategy – surgery may be preferred. If there is associated coarctation of the aorta, balloon dilatation followed by surgical repair of coarctation or both lesions treated by balloon dilation.

Outside Neonatal Period (>1 month): Those with poor LV function balloon angioplasty, decision **not** based on echo gradient. In the presence of normal LV function: intervention if estimated *peak to peak* gradient > 50mmHg, presence of left ventricular hypertrophy, presence of repolarisation abnormalities on ECG, presence of symptoms or if there is exercise induced ischemia.

Postoperative studies

	Basic	Echo	Exercise Treadmill
pre discharge	*	*	
3 months	*	*	
1 year	*	*	
every 1-2 years	*	*	? > 7 years

Early follow up after 2-3 weeks following surgical intervention or if poor function or significant aortic regurgitation at time of discharge following catheter intervention.

8 Aorto-Pulmonary Window

Preoperative studies

Diagnosis is based on echo prior to surgery however a CT angiogram or catheter may be required to image the coronary arteries if the origins are unclear, or if there are concerns regarding pulmonary vascular resistance. It may be possible to close small A-P windows by catheter technique.

Postoperative studies

	Basic	Echo
Pre discharge	*	*
2-3 weeks	*	*
6 months	*	
18 months	*	*
every 2-3 years	*	individualize

9 Atrioventricular Septal Defect

Preoperative studies

All patients should have an ECG, Chest X-ray and Echo prior to surgical intervention plus thyroid function studies in those with Trisomy 21 (usually performed locally).

Cardiac catheterization is reserved for those cases with pulmonary hypertension older than 9 months of age at presentation, or if there are other concerns that cannot be resolved by Echo.

Patients with pulmonary hypertension should be electively repaired in the first 3-5months of life, as delay beyond this is disadvantageous. If there is associated coarctation of the aorta, repair coarctation first (± pulmonary artery banding).

Patients with an ostium primum ASD in isolation (+/- small restrictive ventricular component) should be electively repaired at about 2-4 years of age, unless complicating features such as significant atrioventricular valve regurgitation dictate otherwise.

Postoperative studies

	Basic	Echo
pre discharge	*	*
2-3 weeks	*	*
6 months	*	*
18 months	*	*
Approx. every 2-3 yrs	*	

Individualize ECHO at 18-month visit depending for example on degree of AV valve regurgitation or evidence of subaortic stenosis.

10 Coarctation of the Aorta

Preoperative studies

Primary isolated coarctation: All require Echo. In older patients where echo images are less clear CMR or CT angiography may be required to show clearly the anatomy of the aortic arch.

For those < 6 months surgical repair unless very poor ventricular function or coexisting infection. For those between 6 months and 8 years individualise to either surgery or balloon angioplasty. Above 8 years stent implantation is becoming treatment of choice unless contraindicated by anatomy.

Coarctation with associated critical aortic valve stenosis: Outside spectrum of hypoplastic left heart syndrome treatment depends on severity of each lesion.

Coarctation with associated mitral valve disease: Outside spectrum of hypoplastic left heart syndrome, balloon dilatation or surgery of coarctation if > 6 months (as above), then reassess mitral valve by Echo.

Patients with coarctation and complex intracardiac anatomy: may require catheterization or CT or CMR before surgery.

Coarctation / VSD:

a) Large VSD on echo: repair coarctation + primary VSD repair (or pulmonary artery banding)
b) Small VSD: repair coarctation, no PA band
c) Multiple muscular VSDs or apical VSD on echo: repair coarctation with PA band. Will require catheter before complete repair.

Postintervention

Always obtain a right arm blood pressure and palpate the femoral pulses.

In small infants a lower limb blood pressure should also be performed.

	Basic	Echo	CMR	CT
pre discharge	*	*		6-8 weeks after balloon angioplasty or stenting
2 weeks	*	*		
3 months	*	*		
6 months	*	*		
12 months	*	individualize		
subsequent every 2 years	*	individualize	+ [see comments below]	

Recoarctation

All require echocardiogram, prior to intervention.

In older patients where echo images are less clear CMR or CT angiography may be required to show clearly the anatomy of the aortic arch.

Difficulty in palpating femoral pulses or a cuff blood pressure gradient of > 20mmHg indicates the need for re-intervention. Older children with both native and recurrent coarctation should be considered for stent implantation (cut-off around 30 kg) unless morphology of lesion very discrete.

Post intervention all require MRI in adolescence. The remainder of follow-up is similar to post surgical repair.

11 Coronary artery fistulas

Preoperative investigations

All require echo, followed by cardiac catheterization. CT angiography can also help delineate anatomy. The timing for cardiac catheterization will be dictated by symptoms, age and significance of fistulas (those with dilated main coronaries). Either catheter intervention or surgical ligation may be considered depending on the anatomy of the fistulas.

Small fistulas: As defined by those without main coronary artery enlargement. These may be followed conservatively as many will regress spontaneously with time.

Moderate to large fistulas: These will invariably need treatment either by catheter or surgery, depending upon anatomy.

Postintervention investigations

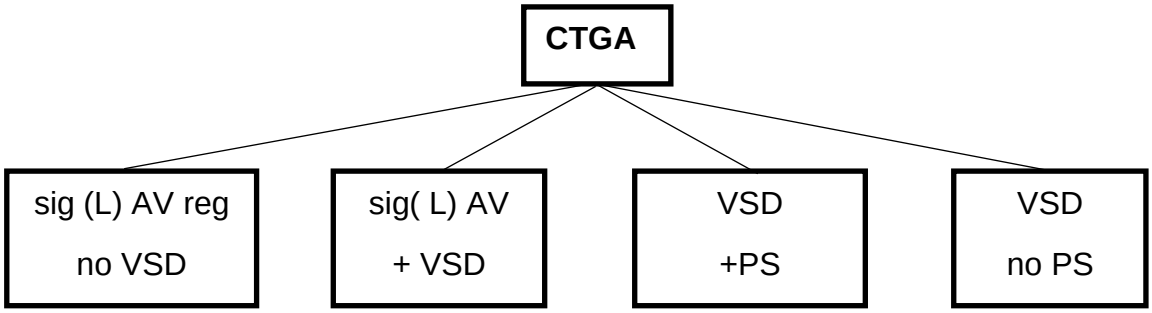
	Basic	Echo	Myocardial perfusion	Holter
pre discharge	*	*		
3 months	*	*		
12 months	*	*		
every 1-2 yrs	*	*	individualize	consider

Anticoagulation may be considered for some cases with residual large redundant proximal coronaries until spontaneous regression occurs.

12 Corrected Transposition (A-V and V-A discordance)

Preoperative studies

All require echocardiography & may require cardiac catheterization prior to complete surgical repair if VSD +/- PS. Holter monitor suggested prior to repair. Surgical management dictated by lesion



Options:

a) cTGA+VSD+PS/PAtresia. Atrial switch plus Rastelli (connect morphologic LV to aorta through VSD + morphologic RV-PA conduit and an atrial switch [Mustard or Senning]). May require initial shunt.
b) cTGA+VSD+noPS: Initial banding and then consider double switch.
c) In some cases, it is appropriate to connect LV to PA if systemic AVV competent (physiological repair).
d) Pulmonary artery banding if significant LAVV regurgitation with TOE mediated adjustment of band in theatre to optimise regurgitation and LV function.
e) No intervention if patient is well balanced (e.g. VSD & PS) with no significant AVV regurgitation

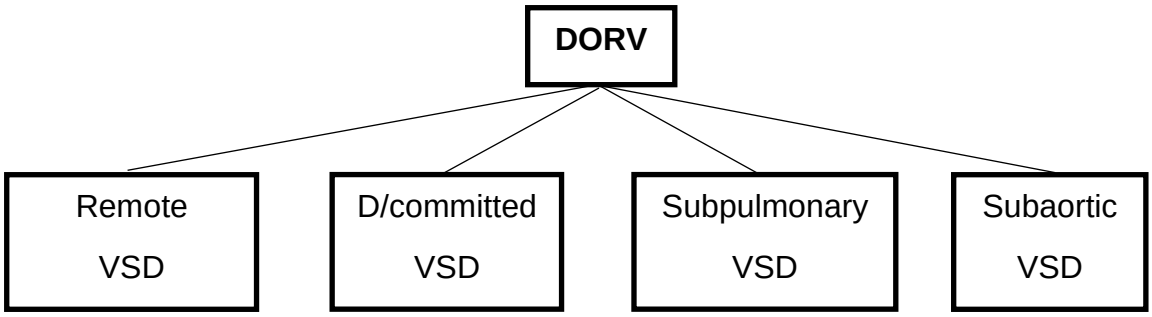
Postoperative studies individualize according to surgical intervention.

	Basic	Echo	Holter	Exercise
pre discharge	*	*		
2 weeks	*	*		
6 months	*	*		
18 months	*	*	*	
subsequent every 1-2 yrs	*		*	? if > 6 years

13 Double Outlet Right Ventricle

Preoperative studies

All require echo study and may require cardiac catheterization +/- TOE prior to complete surgical repair. Initial palliation does not need catheterization.



With significant PS initial palliative shunt with later repair.

With significant additional muscular VSDs: initial PA band and later full repair

With aortic arch obstruction: initial repair with PA band and later full repair.

If initial palliation with subpulmonary VSD, may also require early BAS.

Postoperative studies

	Basic	Echo	MRI & MVO2
pre discharge	*	*	
2-3 weeks	*	*	
6 months	*	*	
18 months	*	*	
subsequent follow-up	*	individualize	adolescent

14 Fontan (Total cavopulmonary Connection (TCPC) pathway for complex lesions

Complex lesions

Definition: This encompasses all cases headed for a TCPC (univentricular A-V connections, and HLHS lesions with significant ventricular hypoplasia, straddling AV valves, remote VSD.... etc)

Preoperative studies

All require echo study prior to surgical intervention. First stage Norwood type procedures (see later), PA bands and BT shunts may be performed without prior cardiac catheterization although

balloon atrial septostomy may be necessary if the ASD is restrictive, particularly with small or absent LAVV.

Postoperative studies

Cases of HLHS will remain as in patients until after the Glenn procedure. Other conditions will be discharged after 1st procedure with close monitoring in clinic and at home. Community nurses will check saturations with frequency at consultant discretion.

Prior to SECOND procedure (bidirectional superior Cavo pulmonary anastomosis) a catheter is required. In some cases, with the branch pulmonary arteries clearly seen on imaging and Doppler assessment indicates a low pulmonary artery pressure it might be possible to proceed without catheter. Consider bidirectional Cavo pulmonary shunt at 3-6 months of age. Additional procedures for subaortic stenosis, unobstructed TAPVC or AVV regurgitation best timed with this procedure.

	Basic	Echo	Oximetry
pre discharge	*	*	* (& Hb)
3 months	*	*	*
6 months	*	individualize	*
18 months	*	*	*

Prior to the THIRD procedure (completion of TCPC) between 2 and 3 years all require catheterization or MRI with SVC pressure measurement. During catheter attention is paid to a) PA pressure & size; b) A-V valve function; c) subaortic stenosis; and d) ventricular function + mass. Based on these findings a decision of either high or low risk TCPC is made, as well as whether fenestration recommended.

Post TCPC

Include oximetry with basic investigations

	Basic	Echo	Holter	Cath	CMR
pre discharge	*	*			
2-3 weeks	*	*			
6 months	*			To close any persistent fenestration	
18 months	*	*			
1-2 years	*	*	*		Older children 2-3 yearly

Anticoagulation as per coagulation guidelines. Renal and hepatic function should be checked every 2 years.

All cases of HLHS will have their data reviewed intermittently in the HLHS specialist JCC MDT meeting. Timing decided by lead cardiologist and surgeon for the patient.

15 Hemitruncus arteriosus

Preoperative studies

All require Echo at time of initial presentation. Cardiac catheterization reserved for case where concerns about pulmonary vascular resistance or if part of a complex lesion.

Postoperative studies

	Basic	Echo	Lung perfusion
pre discharge	*	*	
2-3 weeks	*	*	
6 months	*	*	
18 months	*	*	
Every 2 years	*	individualize	As indicated

16 Hypertrophic Cardiomyopathy

Basic evaluation

Cardiomyopathy screen (see appendix 1) to try and evaluate cause.

Screening of family, involving primary and secondary care teams.

If patient has significant obstruction, pharmacologic manipulation attempted (propranolol and disopyramide). If medical therapy fails and patient symptomatic, consider surgical myectomy.

After initial assessment, if no surgery yearly follow-up with basic, Holter & echocardiogram. If surgery performed follow-up as per sub-aortic stenosis.

17 Hypoplastic Left Heart Syndrome

All require detailed Echo study prior to surgery, to confirm diagnosis and exclude significant associated abnormalities. All patients transferred to the paediatric intensive care unit prior to surgery for pre-op management as HLH pathway. Important to assess ASD size and pressure gradient, degree of A-V valve regurgitation, associated pulmonary venous abnormalities and size of ascending aorta.

Patients counselled: Active treatment is Norwood procedure.

Postoperative studies (Norwood)

	Basic incl. oximetry	Echo	Cath
pre discharge	*	*	
monthly intervals	*	*	
3 months	*	*	*

The post Norwood echo should include an assessment of RV pressure, which is then compared to leg blood pressure, as arm BP readings may be misleading, resulting in missed diagnosis of arch obstruction. Home monitoring required interstage I and II as per liaison nurse protocol.

After the 3-month catheter the patient should follow the pre and post Bidirectional Cavopulmonary shunt/Fontan track. [for example, a bidirectional cavopulmonary shunt at about 3-4 months and TCPC at around 24 months]

18 Interrupted Aortic Arch

Preoperative studies

All require Echo prior to complete repair, cardiac catheterization, CMR or CT angiography if echo assessment unclear.

Postoperative studies

Always obtain a right arm blood pressure and palpate the femoral pulses.

In small infants a lower limb blood pressure should also be performed.

	Basic	Echo	Holter
pre discharge	*	*	
2-3 weeks	*	*	
3 months	*	*	
6 months	*	*	
1 year	*	*	*if previous concerns of arrythmia
every 1-2 years	*	individualize	

MRI or CT of arch if obstruction is suspected at follow up.

Baseline Holter, MRI and exercise testing at 10 years, 14years and transfer to ACHD care.

Consider CT if child unable to comply with MRI. GA MRI can be considered.

Screening cholesterol at 10 years of age.

19 Kawasaki Disease

Normal Coronaries (-2<Z<+2)

	Basic (include ECG!)	Echo
at presentation	*	*
1-2 weeks	*	*
4-6 weeks	*	*
6 months	*	*

- No aspirin beyond the initial 6 to 8 weeks
- After 12 months for follow up at discretion of cardiologist.
- If persistently normal coronaries discharge at 12 months

ANYONE WHOSE CORONARIES ARE ABNORMAL at any stage are for referral to the specialist KAWASAKI clinic (led by Prof Fraisse, Prof Di Salvo and ID Kawasaki experts from ST Mary's Hospitals including Jethro Herberg). Below is a guideline but please note care is individualized in the Kawasaki clinic and at discretion of experts leading the clinic.

Ectasia / Dilatation (2< Z<2.5)

	Basic (include ECG!)	Echo
at presentation	*	*
1-2weeks	*	*
6 weeks	*	*
6 months	*	*
yearly if persists	*	*

- Low dose aspirin (3-5mg/kg/day) until coronaries in normal range
- Discharge when coronaries are within normal range

Aneurysms (Z>2.5)

Echo at least 2/per week until the luminal dimensions stop progressing. And 1/week in the first 45 days in the present of expanding large or giant aneurysm to detect coronary thrombosis. Then monthly until the 3rd month.

See AHA guidelines for further details.

1) Small coronary artery aneurysm (Z = 2.5 – 5.0)

- *Current or persistent*
- *Regressed to normal or dilatation only*

- Assess 6months and then annually
- Consider CT and MRI every 2-5years
- Assessment for inducible myocardial ischemia with Stress Echo/ MRI every 1-3 years
- Long term antiplatelet therapy, until the aneurysms regress
- No restriction to physical activity

2) Medium coronary artery aneurysm (Z = 5.0 – 10.0)

- *Current or persistent*
- *Regressed to small aneurysms*

- Assess at 3 months, 6months and 12 months, then annually
- Coronary CT can be considered and then reconsider CT or MRI every 1-3 years
- Assessment for inducible myocardial ischemia with Stress Echo every 1-3 years
- Long term antiplatelet therapy
- Consult with Anticoagulation service, as increased anticoagulation may be considered (Warfarin/LMWH)
- No contact sports. No other restriction to physical activity.
- Females: reproductive counselling (warfarin risk)

3) Giant coronary aneurysms (Z >10)

- *Multiple complex aneurysms without obstruction, current or persistent*
- *Regressed to medium aneurysm*
- *Regressed to small aneurysm*
- *Regressed to normal or dilatation*

- Assess at 3moths, 6months, 9months and 12 months, then every 6 months
- Consider CT at 6months and then reconsider CT or MRI every 1-3 years
- Assessment for inducible myocardial ischemia yearly with non-invasive stress testing (stress echo or exercise test): Coronary CT-angiogram if myocardial ischemia demonstrated by stress test.
- Cardiac catheterization including coronary CT angiogram 6 -12 months in case of suspected or confirmed myocardial ischemia (ECG, imaging)
- Long term antiplatelet therapy

- Consult with Anticoagulation service, as increased anticoagulation may be considered (warfarin/LMWH)
- No contact sports. No other restrictions of physical activity.
- Females: reproductive counselling (warfarin risk)
- CMC (Coordinate My Care, with directives to the ambulance) +ESP (Emergency Care Plan) +SCP (School Care Plan) if age appropriate.

4) Patients with coronary artery obstructions (calcifications/clots)

- Long term antiplatelet therapy
- Warfarin, target INR 2 – 3.5
- Restricted physical activity as guided by stress test. No contact sports
- KD clinic every 6monyhs
- Myocardial perfusion imaging including stress test (patient >10 years) annually
- Coronary CT if myocardial ischemia demonstrated by stress test.
- For acute chest pain, follow ACS- KD guidelines
- CMC+ECP+SCP (if age appropriate)

20 Patent Arterial Duct

Preoperative studies

With typical findings verified by Echo, occlusion by catheter is first line management at Royal Brompton. Under rare circumstances surgery may be required. If not in failure then manage medically until 5 Kg, for device occlusion. Tiny silent ducts do not require treatment.

Postop/occlusion studies

	Basic	Echo
Occlusion	3/12	3/12
Surgery	3/12	3/12

Discharge surgical patients after one follow-up visit. Discharge device patients after 2 years.

If a residual leak present review yearly.

If clinically silent patent duct detected and patient over 2 year of age, discharge to PEC/GP to listen in 2-3 years and refer if murmur develops or alternatively offer follow up in 5 years.

21 Pulmonary atresia and ventricular septal defect

Preoperative studies

If no aortopulmonary collaterals may not need a catheter if anatomy is clear. If aortopulmonary collaterals are suspected, all need a catheter or CT prior to surgery. NB blood products and conotruncal lesions (see above).

Pulmonary Arteries (confluent + PDA)

<3.0 mm PA's	>3.0 mms PA's
Outflow tract patch	BT shunt, later repair ideally when adequate conduit can be accepted

Pulmonary Arteries (non confluent, blood supply varied)

Individualize, RV outflow tract patch and staged unifocalization with occlusion collaterals causing dual supply with eventual RV-PA conduit ± VSD closure. RV-PA conduits will need subsequent replacement, timing depending upon the amount of conduit stenosis, regurgitation and right ventricular size and function.

Postoperative studies (Repaired)

	Basic	Echo	Cath	CMR& MVO2
pre discharge	*	*		
2-3 weeks	*	*		
6 months	*	*		
12 months	*	*		
18 months	*	*		
1-2 yearly	*	*	If concern	In adolescence

Investigations may be varied as to whether, angioplasty, stenting or embolization are necessary.

22 Pulmonary Valve Stenosis

Preoperative

All require Echo prior to intervention. If classic dysplastic valve take care to exclude associated pathology on the left side, e.g. Noonan's syndrome.

If the peak instantaneous gradient is greater than 60mmHg, then a balloon dilatation should be attempted.

For those with gradients that do not require intervention, 6 monthly follow-up with echo and ECG for those ≤ 2 years of age, 1-2 yearly ≥ 2years. Consider 3yrly appointments and then discharge from follow-up of adolescents whose Doppler gradient is less than 25 mmHg.

Patients with a dysplastic valve should have an attempted dilatation, however surgery is usually necessary.

In the first week of life balloon angioplasty should be performed on Prostaglandins, even if the right ventricle is small.

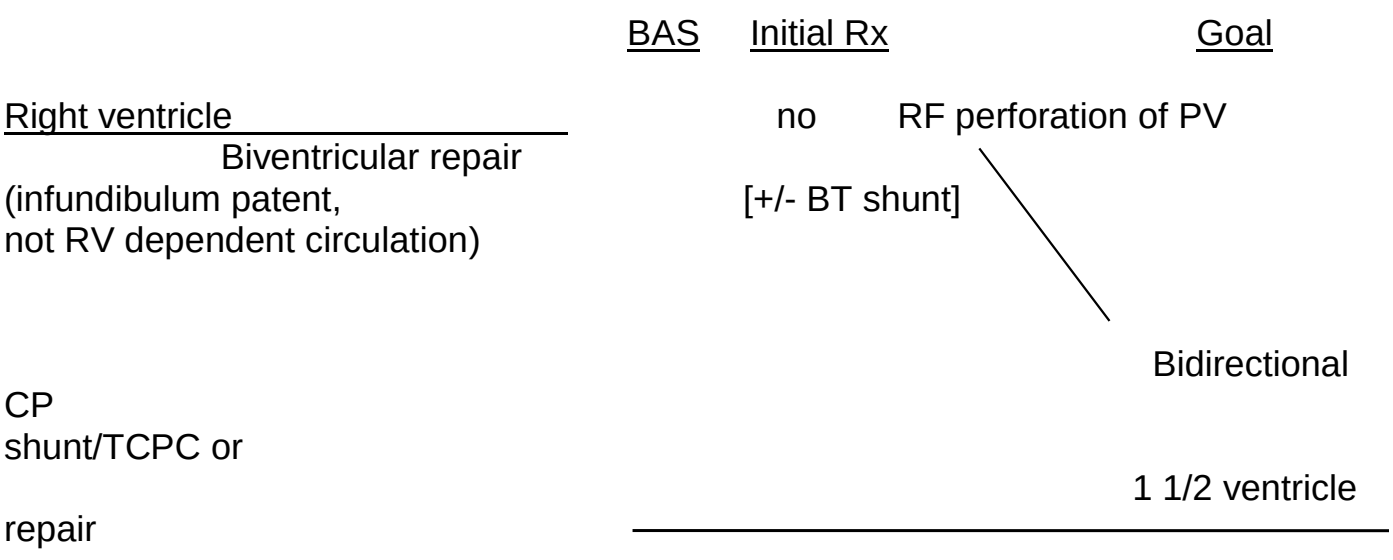
Postintervention

	Basic	Echo	CMR
3 months	*	*	
1-2 yearly	*	*	If significant pulmonary regurgitation

23 Pulmonary Atresia with Intact Ventricular Septum

Preoperative studies

All patients require Echo prior to intervention. Cardiac catheterization may be indicated if there is suspicion of a RV dependent coronary circulation.



Postoperative studies

For those on a Fontan track, see pre Fontan assessment.

For those for a biventricular repair, without a BT shunt, follow as per isolated pulmonary valve stenosis.

For those with a borderline RV size and a BT shunt, Echo and catheterization prior to deciding if shunt and/or ASD can be closed surgically or by catheter.

24 Secundum Atrial Septal Defect

Preoperative studies

Only Echo with classical features, yearly review, with repeat echo prior to closure.

Patients with a PFO (atrial communication < 3 mm) and no right heart volume overload should be discharged.

In cases where the ASD size or location is uncertain a transesophageal echocardiographic study might be required prior to intervention.

If the pulmonary veins can not be identified, a cardiac catheter/ CMR or CT angiogram may be required to show where the pulmonary veins connect.

Postoperative studies

	Basic	Echo	Holter
pre-discharge	*	*	
2-3 weeks	*	*	
6 months	*		
18 months	*	*	
every 5 years	*	*	* for Sinus venosus

For device closure first follow-up at 3 months and then 1-2 yearly, should remain on low dose aspirin until first follow-up visit. Discharge patients with secundum ASDs 5 years after treatment. Sinus venosus ASDs require continuing follow-up 2-3 yrly.

25 Fibromuscular Subaortic Stenosis

Preoperative studies

All require Echo assessment prior to surgical repair. If the *mean* gradient is 40-50 mmHg, then repair, especially in the presence of aortic regurgitation and left ventricular hypertrophy

All require preoperative serial Echo assessment yearly.

If there is an aortic commissural abnormality and/or aortic regurgitation, then earlier repair may be necessary.

Postoperative studies

Note that this lesion can recur

	Basic	Echo
pre-discharge	*	*
2-3 weeks	*	*
6 months	*	*
18 months	*	*
every 2 to 3 years	*	*

26 Tetralogy of Fallot

Preoperative studies

All require Echo prior to intervention. If undergoing a shunt procedure or repair then catheterization is unnecessary if the anatomy is clear. If there is concern of aortopulmonary collateral arteries, branch pulmonary artery stenosis or anomalous LAD across the right ventricular outflow tract, then further imaging (cardiac catheterization or CT angiography) is required. At catheterisation, “down the barrel” view for coronary arteries.

Require catheter or CT angiography prior to a repair if previous shunt has been performed. NB blood products and conotruncal lesions (see above).

Indications for repair

Elective repair: between 3-9 months irrespective of whether or not have had a shunt. If symptomatic 3- 6 months repair unless small pulmonary arteries or severe spelling.

Postoperative studies (repair)

	Basic	Echo	Holter	MRI	MVO2
pre-discharge	*	*			
2-3 weeks	*	*			
6 months	*	*			
18 months	*	*			
every 2-3 yrs	*	individualise	*	> 10 years	at > 10 yrs

Catheter/CMR during follow-up if suspicion branch pulmonary artery stenosis or prior to pulmonary valve replacement.

Most patients will require pulmonary valve replacement in adolescence/ early adulthood.

27 Complete transposition of the great arteries (intact ventricular septum)

Preoperative studies

All patients require an Echo. If intracardiac anatomy and coronary pattern is clear, then balloon atrial septostomy can be performed if required in the PICU or NICU.

If there is coarctation, leave on prostaglandin prior to repair of both lesions. Infants beyond 6 weeks of age may require 2-stage repair to prepare LV before switch.

Postoperative studies

	Basic	Echo	Catheter & CMR	Holter
pre discharge	*	*		
2-3 weeks	*			
6 months	*	*		* 6-12 months if previous arrythmia
18 months	*	*		
every 2 years	*	*	Individualise	• Age 4yrs, 10 & 14yrs and at transfer to ACHD team

Consider Lipid panel age 10 yrs.

Catheter or CMR to look for branch PS or if there are concerns on echo.

Exercise test and CMR myocardial perfusion imaging at 14 years of age, sooner if concern of myocardial ischaemia

28 Truncus Arteriosus

Preoperative studies

All patients require an ECG & Echo prior to surgical repair. Cardiac catheterization reserved for cases where anatomy unclear or there is concern of the pulmonary vascular resistance. Genetic

testing for 22q11 deletion should be performed. If no result available presume diagnosis and use irradiated blood products and conotruncal lesions.

Early repair is recommended unless there are complicating lesions.

Postoperative studies

	Basic	Echo	MVO2 & CMR	Holter
pre discharge	*	*		
2-3 weeks	*	*		
6 months	*	*		If previous arrhythmia concerns .to be done at 1 yr of age*
18 months	*	*		
every 2-3 years	*	*	> 10 years age	If previous arrhythmia concerns .to be done at 4 yr of age* Repeat Holter at 10 & 14 yrs of age.

Cardiac catheter or CT/MRI if concern regarding branch pulmonary arteries on echo. Perfusion scan may be required if echo suggests asymmetric pulmonary blood flow.

The RV-PA conduit will need replacement or intervention if significant stenosis/ regurgitation

Holter before conduit replacement.

Screening cholesterol aged 10 yrs.

Holter, Exercise test, MRI to be considered at 10 & 14 years of age and at transfer to ACHD care.

29 Ventricular Septal Defect

Preoperative studies

Patients who require surgery need a detailed Echo prior to surgery. Associated lesions such as RV muscle bundles, subaortic stenosis and aortic valve prolapse should be excluded or confirmed. Catheterization if concern regarding pulmonary resistance or if associated lesions are not adequately imaged.

Those with significant multiple muscular VSDs should have a PA band. Those with a perimembranous VSD and small associated muscular VSDs should be repaired and may or may not require a pre-op catheterization, depending on the size and location of the associated muscular defects.

VSD does not always require intervention, follow-up individualized. For those with perimembranous VSDs repeat echo recommended 2-3 yearly to exclude the development of associated lesions.

Patients with a small restrictive muscular VSD with no evidence of significant left heart dilatation can be discharged back to Paediatrician or GP age 6-12 months of age with advice to re-refer after 5 years if murmur still present.

Postoperative studies

	Basic	Echo
pre discharge	*	*
2-3 weeks	*	*
6 months	*	
18 months	*	+
5 years	*	*

Aortic regurgitation is an indication for surgical closure. If mild, child should be follow-ed up yearly to look for progression.

Discharge at 5 years post surgery if no concerns and normal ECG.

Patients with VSD device closure should not be discharged currently, but followed-up every 1-2 2 years

30 Authors

Written by AG Magee June 2009. 1st Revision by N Naqvi, Michael Rigby & Rodney Franklin February 2011, 2nd Revision N Naqvi – circulated and checked by cardiology consultant group May 2019.

31 References

1. Wernovsky G, Lihn SL, Olen MM. Creating a lesion-specific "roadmap" for ambulatory care following surgery for complex congenital cardiac disease. Cardiol Young. 2017;27(4):648-62.
2. Hass NA, Schirmer KR et al. Guidelines for the Management Of Congenital Heart Diseases In Childhood and Adolescence. Cardiol Young. 2017;27(Suppl 3) S1-105.