Trust Guideline for Cardiology Referral Pathway; Paediatrics

A Clinical Guideline

For use in:	NICU, The Jenny Lind Department of Paediatrics, Blakeney ward, A&E
By:	Paediatric Medical, Surgical and Neonatal doctors
For:	Neonates and Children with congenital cardiac lesions
Division responsible for document:	Paediatrics
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If Yes - does the strategy/policy deviate from the recommendations of NICE? If so why?	N/A

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Version and Document Control:

Version Number	Date of Update	Change Description	Author
6	09/03/2021	Referral table added as appendix	Rahul Roy

This is a Controlled Document

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Glossary

GD	Graham Derrick	SVT	Supraventricular Tachycardia
RR	Rahul Roy	QTc	Correct QT
AS	Aravind Shastri	WPW	Wolff-Parkinson-White
FM	Florian Moenkemeyer	ECG	Electrocardiogram
PEC	Paediatrician with Expertise in Cardiology	EEG	Electroencephalogram
VSD	Ventricular septal Defect	MR	Mitral Regurgitation
ASD	Atrial Septal Defect	TR	Tricuspid Regurgitation
PS	Pulmonary stenosis	MVP	Mitral Valve Prolapse
AoS	Aortic Stenosis	ICC	Inherited Cardiac Condition

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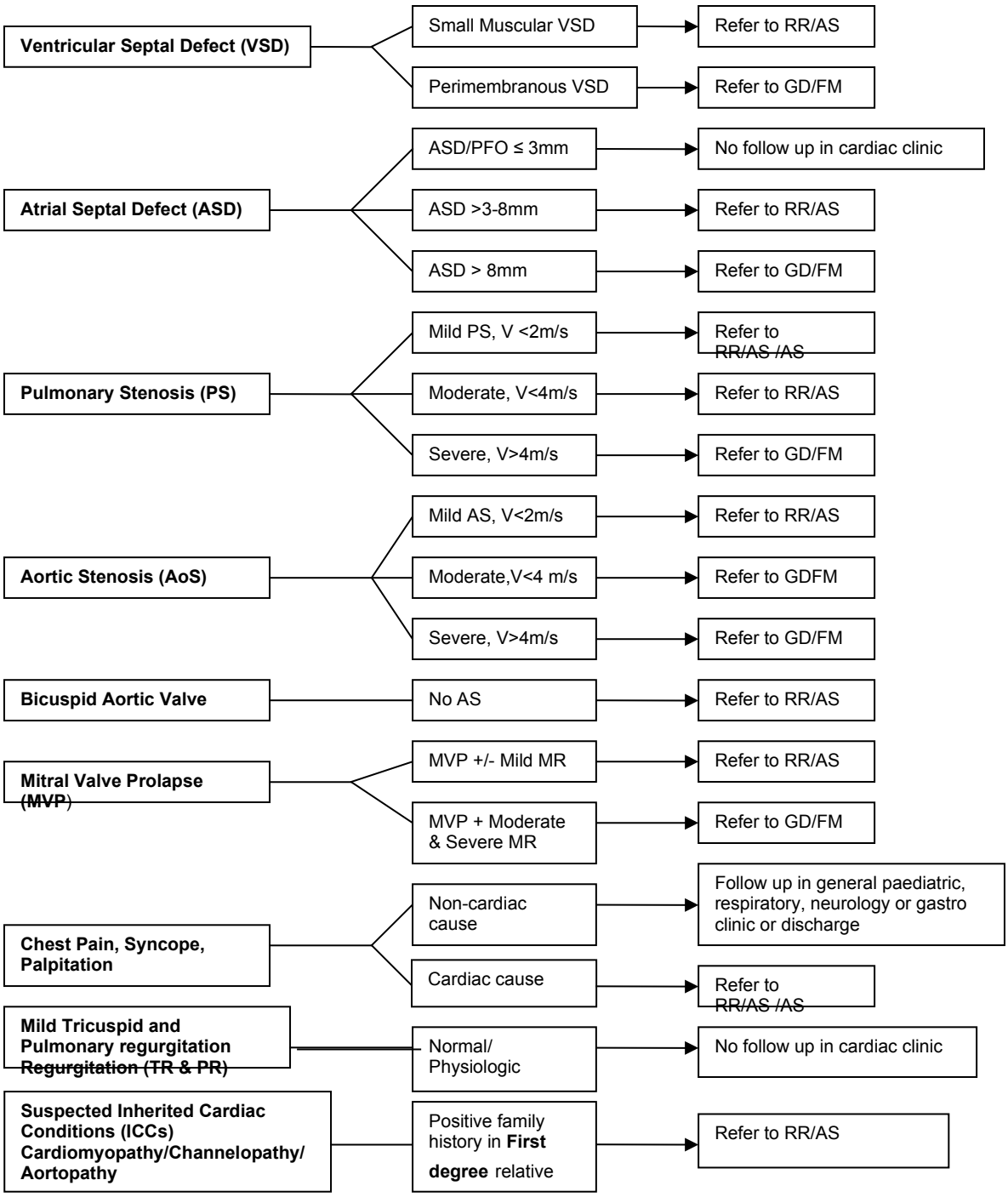
Quick reference guideline

Paediatric Cardiology clinics: referral pathway

Paediatric Cardiac lesions are to be referred for assessment to Dr Rahul Roy (PEC), Dr or Aravind Shastri (PEC) locally. Tertiary outreach Paediatric Cardiology clinic is run jointly with Dr Graham Derrick (GD, Consultant Paediatric Cardiologist) or Dr Florian Moenkemeyer (FM). Please follow the algorithm below to refer to the appropriate Consultant.

Any request for echocardiogram has to be discussed and approved by the consultant in charge of the patient.



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Objective

- To define the services offered by the local Paediatrician with expertise in Cardiology (PEC).
- To define the services offered in the outreach paediatrician cardiology clinics run jointly with the tertiary centre Paediatric Cardiologist.

Rationale

The provision of paediatric cardiology services in a hub and spoke configuration is widely accepted. The workload undertaken in the outreach paediatric cardiology clinics by the Paediatric Cardiologist from the tertiary centre is continuing to increase. There has been an increase in the number of referral of children with asymptomatic murmur for evaluation by echocardiography.

This is in part due to increased detection of murmurs by general practitioners and paediatricians and the need for reassurance of cardiac normality.

The concept of joint outreach paediatric cardiology clinics where a paediatric cardiologist works closely with a paediatrician with an interest in paediatric cardiology would ensure efficient use of the time of the paediatric cardiologist so that the more needy patients can be easily fitted into these clinics.

These joint clinics will ensure that The British Congenital Cardiac Association (BCCA), Royal College of Paediatrics and Child Health (RCPCH) guidance and recommendation of the Department of Health document The Paediatric and Congenital Cardiac Services Review (PCCSR) for Paediatrician with expertise in cardiology (PEC) and issues of clinical governance are fulfilled. This local paediatrician delivered cardiology service will endeavour to make the patient journey from referral to specialist opinion and assessment a seamless pathway.

Broad recommendations

The incidence of congenital heart disease is 8 in 1000 live-born babies and this has remained consistent over several decades. The prevalence will continue to rise, as treatments continue to become more successful. Any child with a cardiac abnormality will be seen or discussed with the Paediatric Cardiologist and would have a clear plan for follow up, jointly agreed by the cardiologist and local paediatrician. Clinical governance issues related to maintenance of expertise, continuous professional development, audit of practice, including workload and patient referral pattern will be carried out.

Also see this guideline need to be cross referenced to the O & G guideline entitled "Trust Guideline for Referral when a Fetal Abnormality is detected" – Trustdocs ID: 882 (guideline number AO16).

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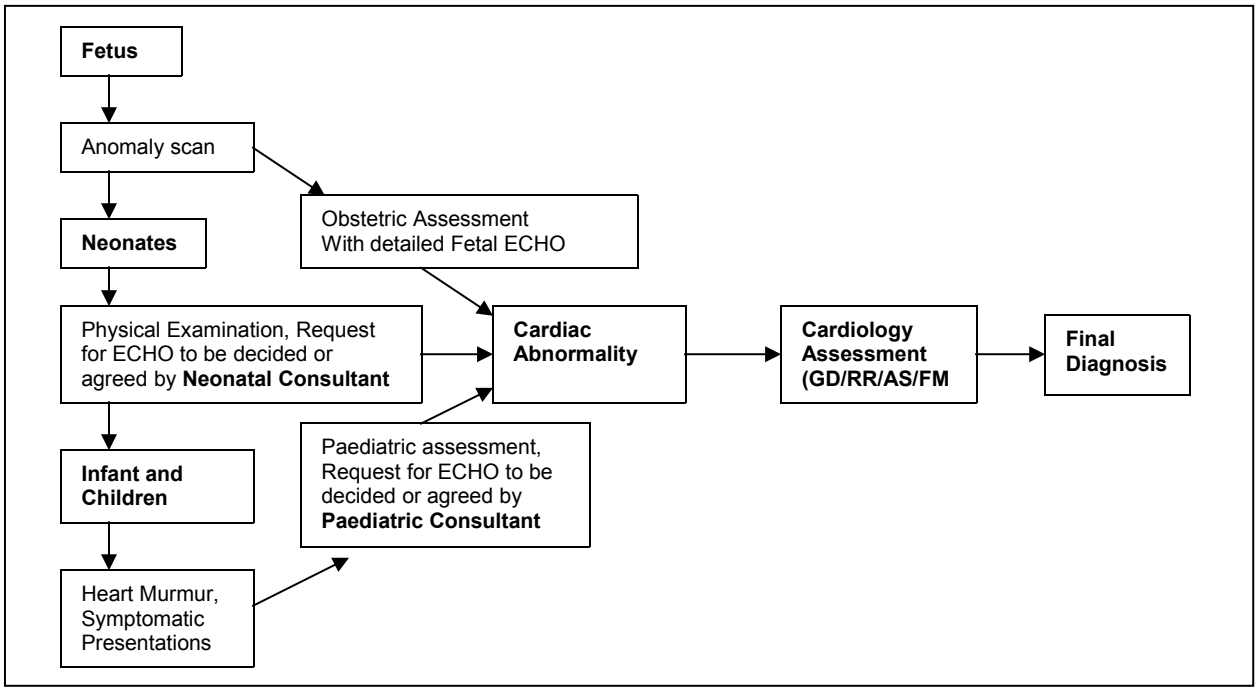
Care Pathway- Identification to Diagnosis



Useful referral details

Urgent assessment (immediate to 24 hrs) - Refer to AS/RR/Cardiac physiologist 0830 to 1700 – contact via switchboard/secretary

Out of hours - There is no dedicated on-call service but discussion with RR/AS/cardiac physiologist can be attempted first; confirmed/strong suspicion of CHD to be discussed with GOSH

Routine referrals - via clinic referral letter or intranet online referral form

GP Out-patient referrals – Referrals for murmurs in children more than 6 months of age and referrals for chest pains, syncope and palpitations can be seen in general paediatric OP clinic (GENJL) and then referred to PEC clinic if ongoing symptoms or to tertiary clinic if any definitive cardiac diagnosis

12 lead ECG - This can be done by trained paediatric nurse or obtained via cardiology department after referral on ICE

Holter monitoring – Please first decide if it is needed (See detailed explanation of various symptoms as below); Discuss with named consultant or AS/RR/GOSH team if any doubts. If any abnormalities, d/w AS/RR/GOSH

Exercise test - This service is not routinely available at NNUH. In exceptional cases, under supervision of paediatric registrar or after d/w AS/RR – this could be undertaken at NNUH.

Innocent Heart Murmur

More the 80% of children have innocent murmurs of one type or another sometime during childhood. Only 0.8 -1% has structural congenital heart disease. When one or more of the following are present, the murmur is more likely pathologic and requires cardiac consultation and/or echocardiogram:

- Diastolic murmur
- Loud Systolic murmur i.e. grade 3/6 or with a thrill, long in duration and transmits well to other part of the chest.
- Abnormal heart sounds, systolic click or opening snap.
- Central Cyanosis.
- Abnormal peripheral pulses.
- Continuous murmur that cannot be suppressed.
- Abnormal ECG.

Small Midmuscular & apical Ventricular Septal Defect (VSD)

VSD is the most common form of congenital heart defect and accounts for 20% of congenital heart disease. A VSD can be classified as a perimembranous or a muscular defect. Muscular VSD are frequently multiple. Patient with small muscular VSD are asymptomatic. A pansystolic murmur is best audible at the lower left sternal border, occasionally the murmur is early systolic. Spontaneous closure occurs frequently in small defects. Small muscular VSDs will be followed up 1-3 yearly in RR clinic till it undergoes spontaneous closure.

Secundum Atrial Septal Defect (ASD)

Three types of ASDs exist: secundum, primum and sinus venosus defect. Most common type of ASD is the secundum defect which accounts for 5-10% of all congenital heart defects. Spontaneous closure is common in the first 2-3 years.

ASD/PFO ≤ 3mm in size spontaneous closure occurs in majority of cases and requires no follow up.

ASD > 3mm to 8mm less likely to close spontaneously but can get smaller. Follow up every 6-12 months in RR/AS clinic.

ASD ≥ 8mm rarely closes spontaneously.

Primum ASD and sinus venosus ASD – to be seen GD/FM clinic

Most children with an ASD remain asymptomatic. Those who become symptomatic with recurrent lower respiratory tract infections or have a large ASD can be closed by trans catheter occlusion device/surgery around 3-5 years age. They would be referred GD/FM.

Pulmonary Stenosis (PS)

PS occurs in 8% to 12% of all congenital heart defects. PS may be valvar (90%), subvalvar or supravalvar. Pulmonary valve leaflets may be thin or thickened with restricted systolic motion.

Classification of stenosis:

Velocity < 2 m/s – very mild PS
2-3 m/s – Mild PS
3 to < 4 m/s – moderate PS
> 4 m/s – severe PS

Children with mild PS are completely asymptomatic. Mild PS is usually not progressive but may improve over time. Mild to moderate PS will be followed up in RR/AS clinic every 4 -18 months.

Aortic Stenosis (AoS)

AS occurs in 3% to 6% of all congenital heart defects. AS may be valvar (70%), subvalvar or supravalvar.

Classification of stenosis:

Velocity < 2 m/s – mild AS
< 4 m/s – moderate AS
> 4 m/s – severe AS

Most children with mild to moderate AS are asymptomatic. Chest pain, syncope, and even sudden death may occur in children with severe AS. AS can progressively worsen with time. Mild AS will be followed up every 6 -18months in RR/AS clinic.

Bicuspid Aortic valve

Valvar AS may be caused by a bicuspid aortic valve with fused commissures hence need follow up every 1 – 3 years in RR/AS clinic.

Mitral Valve Prolapse (MVP)

The mitral valve leaflets are thick and redundant. The posterior leaflet prolapses more commonly than anterior leaflet. Mild Mitral Regurgitation (MR) is occasionally demonstrated. MVP with or without mild MR will be followed up 1-2 yearly in RR/AS clinic.

The majority of patients are asymptomatic but history of non-exertional chest pain, palpitation, and rarely syncope may be elicited.

Symptomatic patients and those with severe MR will be followed up in tertiary clinic.

Tricuspid and Pulmonary regurgitation (TR and PR)

Mild TR without any dilation of the right atrium and right ventricle do not need follow up in cardiac clinic. Mild PR without any dilatation of right ventricle or enlargement of main pulmonary artery also doesn't need follow up in cardiac clinic.

Antenatally diagnosed vascular rings (Double aortic arch and Right arch with Aberrant Left subclavian artery)

No other extra-cardiac diagnosis – Follow up with AS/RR with one f/u with GD/FM
Other non-cardiac diagnosis needing follow up – F/u with relevant named consultant.

Antenatally diagnosed major CHD

Prematurity < 34 weeks or other important non-cardiac diagnosis - please arrange follow up with named neonatal consultant.

Cardiac diagnosis only - refer to RR/AS for local follow up.

Chronic lung disease of prematurity

Patients needing O2 after 36 weeks of corrected age and are due to be discharged home on Oxygen need to have an echocardiographic assessment prior to discharge for following indications

- Baseline echocardiogram ensuring normal intra-cardiac anatomy and also to see if previously diagnosed conditions like PDA have resolved completely.
- If any evidence of pulmonary hypertension.
- If pulmonary hypertension, to understand the pulmonary venous anatomy in more details.
- If O2 requirement worsening after discharge, repeat ECHO in 4 months; if persistent O2 even at 1 year of age, another echocardiogram can be requested.
- Those needing HHFNC treatment at 36 weeks CGA or significant O2 (> 1L/min) even at 36 weeks CGA will need earlier ECHO (between 35-36 weeks).

Family History of Congenital Heart Disease/Inherited cardiac conditions

- Please Refer to table 1 and key messages at the end of table

Chest pain

Chest pain is relatively common in children but extremely rarely cardiac in origin. Careful history and examination particularly palpation of the chest is essential to exclude musculoskeletal causes e.g costochondritis. Gastrointestinal disorders e.g gastrointestinal reflux and respiratory problems can also cause chest pain.

Chest pain can be associated with structural lesions of the heart, in particular aortic stenosis, aortic dissection (Marfan's syndrome) and mitral valve prolapse. Tachyarrhythmias like Supraventricular Tachycardia (SVT), Ventricular Tachycardia (VT) can cause chest pain along with palpitation and breathlessness. Pericardial disease such as acute pericarditis and post-percardiotomy syndrome can cause chest pain.

Palpitation

Palpitation is one of the most common cardiac symptoms and but poorly correspond to demonstrable abnormalities. However, palpitation may indicate the possible presence of serious cardiac arrhythmias.

Careful and detailed history should be taken. The nature and onset of palpitation, relationship to exertion, associated symptoms, personal drinking habits and family history of sudden death, syncope, or arrhythmias should be enquired.

A routine 12 lead ECG should be taken to exclude prolonged QTc interval, WPW pre-exitation or AV block. A 24 -72 hour Holter monitoring is usually helpful in making a diagnosis of the rhythm if palpitation occurs frequently. When palpitation occurs infrequently, long term event monitoring is indicated. Investigations are helpful for reassuring patients and family if the patient has identified symptoms during the recording period with no evidence of significant cardiac arrhythmias or an AV conduction disturbance identified. This patients and families can be reassured and need no cardiac follow up.

Syncope

Syncope is a common problem in children and adolescents between the ages of 8 and 18 years. Before the age of 6 years, syncope is unusual except in the setting of seizures, breath holding and cardiac arrhythmias. The vast majority of syncope in children and adolescents are benign, resulting from vasovagal episodes, hyperventilation, orthostatic hypotension, and breath holding. Cardiovascular causes of syncope are rare in adolescence, but it is important to be aware of them, as they are potential causes of sudden death. Cardiac causes of syncope include structural lesions, myocardial dysfunction, and arrhythmias, including long QT syndrome.

Syncope in response to loud noise, fright, emotional stress, during exercise, whilst supine, associated with tonic-clonic or abnormal movements and family history of sudden death in young person are strong 'warning bells' from the history.

The key to the diagnosis of syncope is to take a careful and detailed history.

The most important investigation is a 12 lead ECG. The ECG should be inspected for arrhythmias, a long QTc interval, WPW pre-excitation, heart block and abnormalities suggestive of cardiomyopathies. Holter monitoring is usually unhelpful, as symptoms almost never occur in the 24-72 hour period while the monitor is worn. Unless the child has other cardiac signs and symptoms, or any

of the warning bells from history, an echocardiogram will almost certainly be normal. EEG is often performed on children with syncope to 'exclude epilepsy'; this is rarely helpful for even in children with epilepsy as the EEG will usually be normal between attacks.

If there is good history for vasovagal syncope and the 12 lead ECG is normal, usually no further investigation and simple reassurance is all that is usually required. Increased dietary salt and fluid intake can be encouraged and advice on posture (i.e. crossing the legs and folding the arms) when prodromal symptoms are experienced whilst standing can be helpful. Low dose Fludrocortisone can be tried in some cases.

Inherited Cardiac Conditions (ICCs)

Inherited cardiac conditions (ICCs) can affect people of any age and can be life threatening. ICCs are a group of largely monogenic disorders affecting the heart muscle, its conducting system and vasculature. For many families, the first indication there's a problem is when someone dies suddenly with no obvious cause or explanation. Inherited cardiac conditions are caused by a fault (or mutation) in one of more of our genes. If one of the parents has a faulty gene, there's a 50:50 chance the child could inherit it. Where an index case is identified, screening will be offered to first degree relatives, and cascaded to others as deemed necessary on the basis of risk

The most common inherited heart conditions are:

Inherited heart rhythm disturbances, for example:

- Long QT syndrome (LQTS)
- Brugada syndrome
- Catecholaminergic polymorphic ventricular tachycardia (CPVT)

Cardiomyopathies, for example:

- Hypertrophic cardiomyopathy
- Dilated cardiomyopathy
- Restrictive cardiomyopathy
- Arrhythmogenic right ventricular cardiomyopathy

Inherited Arteriopathies, for example:

- Marfan syndrome
- Ehlers-Danlos syndrome
- Loeys-Dietz syndrome

Muscular Dystrophies

- Emery-Drefuss muscular dystrophy
- Myotonic dystrophy

After assessment by the local PEC, all patients with suspected ICCs and/ or at risks of sudden cardiac death will be referred or discussed with GD/FM regarding on going management. Need for referral to the inherited cardiac disease specialist services in Great Ormond Street Hospital, London will be decided by GD/FM.

Clinical audit standards

1. Referral to the Paediatrician cardiology clinic (RR/AS) should strictly comply with the cardiology referral pathway.

Summary of development and consultation process undertaken before registration and dissemination

This guideline was drafted by Dr Rahul Roy on behalf of the Paediatric directorate and modified by Dr Rahul Roy/Aravind Shastri in Oct 2019. During its development it has been circulated and presented to the consultant paediatric cardiologist, neonatologist and paediatrician for comments. The guideline was reviewed by Cardiologist from GOSH Dr Graham Derrick in Sept 2015. It was reviewed again in 2019 where only minor amendments were made.

Distribution list / dissemination method

Trust Intranet

References and source documents

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Appendix 1 - Cardiology Referrals for family history of CHD

CHD in First degree relative or multiple second degree relatives	Fetal medicine(FM) unit ECHO at NNUH	Fetal Cardiologist ECHO at GOSH	Postnatal (NIPE)Examination and Pulse-oximetry	Postnatal ECHO/investigations for baby
VSD/PDA/AVSD needing intervention/surgery	Yes	No Refer if any concerns in FM-scan	As standard	No
ASD needing surgery/intervention	Yes	No	As standard	Routine ECHO in 3 months** (Request Cardiac Physiologist ECHO)
Family h/o Pulmonary stenosis/Aortic stenosis	Yes	No	As standard	No
Family h/o Aortic stenosis related to bicuspid aortic valve	Yes	No	As standard	Refer to AS/RR routinely
Transposition of great vessels	Yes	No	As Standard	No
Tetralogy of Fallot/Truncus	Yes	No	As standard	No
TAPVD	Yes	Yes	As standard	No
Hypoplastic left heart syndrome (HLHS) or Hypoplastic right heart syndrome	Yes	No/discuss if any concerns	Keep baby for 24hrs, check femorals at discharge; Open access to CAU for 4weeks	Request Cardiac physiologist or AS/RR ECHO
Coarctation of Aorta	Yes	No/discuss if any concerns	Keep baby for 24hrs, check femorals at discharge; Open access to CAU for 4weeks	Request Cardiac physiologist or AS/RR ECHO
Cardiomyopathy/Channelopathy in first degree relatives only	Yes	No	As standard; ECG 24 hrs later	Refer to AS/RR as per cardiology pathways
Bicuspid aortic valve/Aortopathy in first degree relative	Yes	No	As Standard	Refer to AS/RR as per cardiology pathways
WPW syndrome in first degree relative	No	No	As standard	Request ECG at 4 weeks and review by named consultant ; if normal discharge Refer only if abnormal
Heart murmurs in family***	No	No	As standard	No

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Key messages

- Every pregnancy gets an initial dating scan and a detailed anomaly scan at 18-20 weeks as per foetal anomaly screening programme
- Any suspicion of CHD in unborn baby at 20 week-anomaly scan/fetal medicine scan gets referred to tertiary fetal specialist echocardiography(GOSH) as per current practice
- If there is a family history of first degree relative **needing cardiac surgery/intervention, the** pregnant lady gets referred to fetal medicine unit for detailed scan by Foetal medicine consultants at NNUH ; if this scan is normal, there is no need to refer newborn baby except for two indications in first degree relative for baby - ASD which needed surgery , hypoplastic left heart/Coarcation of aorta or Aortopathy
- ** ASD cannot be diagnosed in foetal scans and if a first-degree family member needed surgery/intervention – routine echo in 3 months is advised
- *** Heart murmurs without any diagnosed CHD are common normal findings in children/adults this does not warrant any specialist reviews for the baby ; similarly small holes like PFO/VSD which spontaneously closed in parents or siblings do not warrant an echo for the newborn baby
- Cardiomyopathy and Channelopathies like – Long QT syndrome/CPVT/Brugada are rare; if **first degree** relatives of baby (parent/sibling) truly have the condition, please refer to AS/RR clinic. As this screening needs regular reviews, affected second degree relatives(Eg grandparents, maternal uncle etc) for the baby are not an indication for referral as per national screening policy unless it is already proved that multiple second degree relatives are affected by same condition.