NWCHDN Document Control

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Comments:			

Paediatric Cardiology Outpatient Referral Guideline

Date: April 2021

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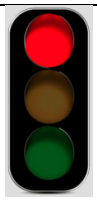
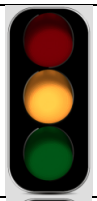
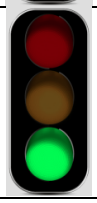
Introduction

This is a guideline for deciding the most appropriate referral pathway for children presenting with common cardiac symptoms and associated diseases. It is applicable for referral to tertiary cardiology and local PEC (paediatrician with expertise) clinics. It is designed to help in the initial assessment and referral process for GPs and paediatricians. In addition, it aims to help clinicians across the network triage and risk stratify planning patient assessments.

Primary assessment should focus on history and clinical assessment. Cardiology tests (holter, exercise) should not be ordered by referring teams where they are not confident in the indication for the test and are not able to analyse the results.

Point of care echocardiography by adult physiology staff or inappropriately trained medical staff should be avoided as this may result in misdiagnosis, delay in referral or inappropriate reassurance or anxiety for the family.

Triage of Referrals

	**RED -** URGENT REVIEW Features in history Symptoms or signs	Cardiology review as inpatient where possible if patient is admitted ***Call to discuss with on call team** Advise patient reviewed in next 2 - 4 weeks (relevant investigations at or prior to review where possible)
	**AMBER** - REQUIRES EARLY REVIEW	Patient to be reviewed in outpatient clinic in next 8 weeks
	**GREEN –** ROUTINE PATIENT REVIEW	Patients are reviewed within the national 18-week framework
	**NO INDICATION FOR PAEDIATRIC CARDIOLOGY REVIEW**	

Please note that all patients that are deemed to be 'urgent' should be discussed on the day with a cardiac registrar or consultant at the Level 1/ 2 centre and plans made for review. Accompanying written referral should be sent confirming key clinical and demographic data

Faxing through referrals marking them 'urgent' does not guarantee timely review.

How to Make a Referral

Tertiary cardiology services in the North West, North Wales and Isle of Man Congenital Heart Disease Operational Delivery Network (NWCHDN) are located at Alder Children's Hospital (Level 1 centre) and Royal Manchester Children's Hospital (Level 2 centre). Many paediatric departments across the network provide Level 3 services provided by a paediatrician with expertise in cardiology (PEC) with many centres hosting joint clinics alongside visiting Level 1 or 2 clinicians. Please note in general that new referrals for those 17 years and above should be directed to adult care (there may be some variation by trust).

Please be aware of your local services and primary referral should usually be sent close to home for review in the first instance. Referrals to Level 1/2 centres in general should be addressed to the paediatric cardiology team rather than an individual to allow for prompt response and triage.

Minimum referral details

Referrals should contain minimum patient demographics of:

1. Patient name
2. Date of birth
3. NHS number
4. GP or Lead consultant responsible for referral/patient

> Pertinent clinical symptoms or signs that are relevant to the presentation should be included in the referral and are denoted by this bullet point throughout this document

> Relevant test results should be attached (genetic reports, holter monitor reports)

Making a Referral to Alder Hey Children's Hospital

All referrals come centrally and are scanned for triage by the consultant team. *Urgent* referrals should also *be discussed with the on-call team*.

Urgent referral contacts

On call Registrar and Consultant available via switch board 0151 228 4811
Registrar on call email Ahc-tr.cardreg@nhs.net

Pathway coordinator contact details
Internal email Cardiology@alderhey.nhs.uk
External email ahc-tr.cardiology@nhs.net

Making a Referral to Royal Manchester Children's Hospital

All referrals are triaged by the consultant team. Urgent referrals should be discussed with the on call team.

Urgent referral contacts
On call Registrar and Consultant available via switch board 0161 276 1234

Paediatric Cardiology secretaries
External email mft.rmchcard@nhs.net

Referral Following a Prenatal Diagnosis

For patients with a prenatal diagnosis of **congenital heart disease (CHD)** the fetal medicine report will clearly document the diagnosis and management plan after birth. See network guidance on delivery planning. Some babies with non-duct dependent lesions may be delivered in local hospitals but tend to warrant early evaluation. This should be discussed with the tertiary centre and a plan for review should be clear prior to hospital discharge. Where there is PEC cover then a local echocardiogram may be performed to confirm the diagnosis and discussed with the tertiary centre.

Please note that in some cases there may be **diagnostic uncertainty** meaning that a complete echocardiogram is advised prior to hospital discharge.

Patients diagnosed with a **right aortic arch** prenatally require standard paediatric checks at birth. Acute airway compromise (as a result of vascular ring) is rare in the first few days. Follow up review should be arranged with paediatric cardiology (for minimum of one assessment) and general paediatric follow-up is advised for the first 2 years to look for symptoms and signs of a vascular ring.

Following a **prenatal diagnosis of arrhythmia** detailed evaluation is required after birth. Maternal medication given prenatally should be documented and cord blood anti-arrhythmic levels should be sent. Baseline ECG should be performed and discussed with cardiology, plans for review should be made prior to discharge from hospital.

Family History of Congenital Heart Disease

The vast majority of screening for congenital heart disease is now done prenatally and it is recommended where either parent has CHD or a previous pregnancy or sibling is affected that a fetal echocardiogram is offered.

The 20-week scan is now robust in examining all areas of the heart, therefore in asymptomatic patients with a normal clinical examination **we do not recommend postnatal cardiac review in most cases**. In *exceptional* circumstances where there has been significant neonatal morbidity or mortality as a result of missed CHD then children may be reviewed for parental reassurance. Please discuss with the cardiology team before making an offer to the parents.

There are some forms of CHD which are difficult to identify prenatally and may be inherited. We would therefore recommend referral for family history (1st degree relative) of:

- Bicuspid aortic valve (anatomical or functional)
- Hypoplastic left heart (to screen for bicuspid aortic valve)
- Laterality disturbance (left or right atrial isomerism)

Ongoing Referral Following an Abnormality Identified on Local or Neonatal Echocardiography or Investigation

All patients identified with CHD should be reviewed on a least one occasion by a paediatric cardiologist. Many patients identified with minor CHD will be suitable for ongoing review in the **PEC clinic**, these would include:

- Restrictive ventricular septal defect (normal RV pressures)
- Small – moderate atrial septal defects
- Small – moderate patent ductus arteriosus
- Mild – moderate pulmonary valve stenosis

Changes or concerns should be discussed with the visiting cardiologist or Level 1 / 2 centre.

Note that a **patent foramen ovale (PFO)** or small atrial communication < 3mm is a normal finding (found in 1 in 4 adults) and observed more commonly in neonates. Most will resolve spontaneously. There is no indication for intervention or lifestyle restriction and so ongoing referral or assessment is not indicated in this group.

Patients with **abnormal chest x-ray** findings suggesting cardiovascular disease should have a complete cardiovascular examination with measurement of saturations and blood pressure before discussion with the cardiology team.

Concerns about **abnormality of resting ECG** should be discussed with cardiology at the Level 1 or 2 centre. Please note we do not accept faxed ECG recordings as the quality is not sufficient to allow remote review. ECGs should be scanned or photographed with a high-resolution camera and sent to the on-call team by confidential e-mail.

Family History of Inherited Cardiac Condition

There is further guidance on this group of patients regarding referral, surveillance and investigation. Please note that across the network we only recommend screening for **first degree relatives**. Screening should be offered in a cascade fashion beginning with first degree relatives of the index case. Cardiologists work closely with clinical geneticists across the network to offer a comprehensive service to these families.

It is vital that where possible the **family history is clearly documented** in the referral:

- Name of first degree relative and date of birth;
- NHS number of family member;
- Send diagnostic details;
- Name of cardiologist/geneticist that provides their follow-up;
- Whether the patient been referred to clinical genetics;
- Ask permission to access medical records of affected individual.

For families who are seeking screening without a first-degree family member then sometimes this can be provided by charitable organisations, such as CRY. https://www.c-r-y.org.uk/screening/

Family History of Premature Cardiovascular Disease

Where there is a family history of premature myocardial infarction in a first degree relative, initial screening of a lipid profile is recommended to screen for familial hypercholesterolemia. Abnormal findings should prompt referral to paediatric metabolic medicine.

For patients with abnormal results within Wales, these patients should be referred to the all-Wales FH service. www.cavuhb.nhs.wales/our-services/all-wales-familial-hypercholesterolaemia-service/

Murmur

Murmurs are common. 80% of children have innocent murmurs of one type or another at some point of these only 1% have CHD. Patients with a clinically innocent murmur during an intercurrent infection should be brought back for repeat auscultation. Referral for cardiac review is recommended in those with a persisting murmur though > 99% of asymptomatic murmurs in childhood will be innocent.

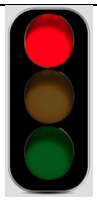
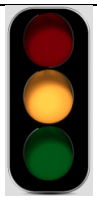
Please note that innocent murmurs will come and go and a child who has previously assessed and diagnosed with an innocent murmur does not require re-referral should a clinically innocent murmur be heard at a later date.

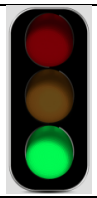
Referrer should examine the child and particularly document in the referral:

- Clinical assessment innocent vs pathological;
- Any associated symptoms or signs e.g breathlessness, difficulty feeding or failure to thrive (FTT);
- Any family history of CHD.

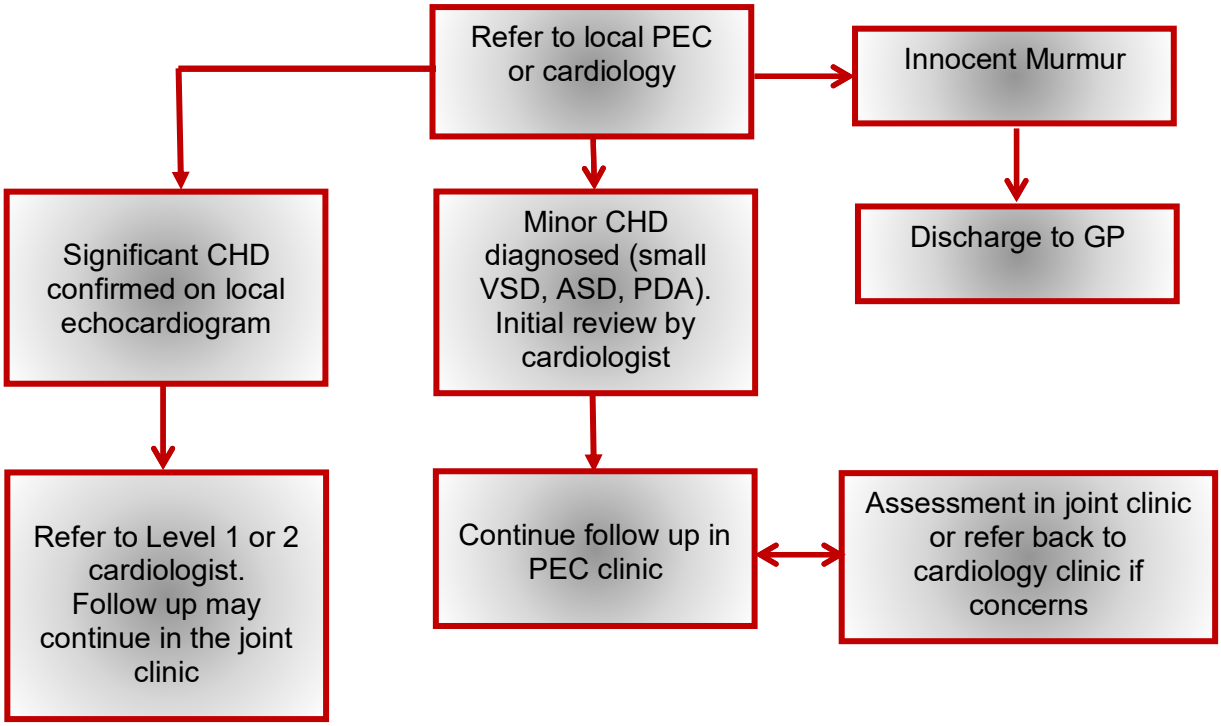
If the patient is assessed in a paediatric setting, then other tests should be performed (where available) prior to cardiac review to evaluate and triage referrals appropriately

- **BP**
- **ECG**
- **Oxygen saturations** (pre and post ductal saturations in neonates)

	Signs of congestive heart failure o respiratory distress, o poor peripheral pulses o decreased urine output o oedema, and/or hepatomegaly Signs of central cyanosis Symptoms/ signs of endocarditis	Cardiology or PEC review as inpatient where possible if patient is admitted *Call to discuss with on call team Aim for review in next 24 – 48 hours
	o failure to thrive o feeding difficulty o neonate / infant < 6 months o signs consistent with pathological murmur o abnormal BP and/or ECG	Patient to be reviewed in outpatient clinic in next 8 weeks

	Clinically innocent murmur o short, soft, systolic o varies with posture variable o no evidence of systemic disease	Patients are reviewed within the national 18-week framework
	o clinically innocent murmur during illness that resolves on repeat auscultation o persistent or intermittent innocent murmur previously assessed in the paediatric cardiology clinic	

Flow Chart for Assessment of Patients with a Murmur



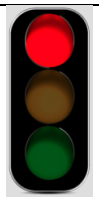
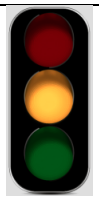
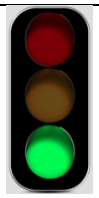
Palpitations

Palpitations can be a commonly described in the paediatric age group. This symptom poorly corresponds with a significant rhythm abnormality. A careful history should be taken to pick up findings that may indicate the presence of a significant cardiac arrhythmia.

Referrer should examine the child and particularly document in the referral:

- Relationship to exertion;
- Any associated symptoms such as collapse or chest pain;
- Family history of sudden cardiac death, inherited cardiac condition, unexplained collapse or arrhythmia.

Referrals from hospital settings should include an **ECG** where possible

	• exertional palpitation with syncope • documented ventricular arrhythmia • patient with known channelopathy or cardiomyopathy	Cardiology review as inpatient where possible if patient is admitted *Call to discuss with on call team Aim for review in next 24 – 48 hours ETT at first review where possible
	• palpitation on exercise • abnormal ECG • FH channelopathy • FH sudden cardiac death/ PPM or ICD < 50yrs • FH cardiomyopathy • Documented supraventricular arrhythmia	Patient to be reviewed in cardiology outpatient clinic in next 8 weeks
	• Recurrent palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG • Palpitations with FH at a young age (before the age of 40 years) of sudden cardiac arrest or death and/or PPM or ICD • PVCs (asymptomatic) in the prenatal or neonatal period	Patients are reviewed within the national 18-week framework by PEC or cardiologist

	• PVCs after the neonatal period (documented >10% on 24 hr tape)	
	• Isolated or resolved palpitations with no other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG • Infrequent SVEs or monomorphic PVCs noted • Sinus bradycardia (asymptomatic) • Sinus arrhythmia	

Peripheral Cyanosis

Evidence of bluish discolouration *around* the mouth (perioral cyanosis), hands or feet particularly when cold is a common finding in young children particularly toddlers. Children are well when this is observed, and this resolves spontaneously or on warming up. In the context of a normal examination and no other concerns in the history this does not warrant any further investigation.

Central Cyanosis

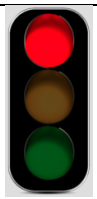
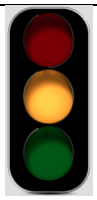
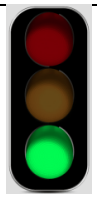
Observation of central cyanosis in children warrants urgent discussion and investigation. Where possible measured oxygen saturations should be checked. Desaturated patients should be discussed urgently with the cardiology on-call team at AH or RMCH. Careful cardiac examination and ECG and chest x-ray should be performed locally.

In neonatal patients pre- and post-ductal saturations should be measured.

Syncope or Vasovagal Symptoms

Vasovagal symptoms and syncope are common in the teenage population. Below the age of 6 years, syncope is unusual except in the setting of seizures, breath holding, reflex anoxic seizures and cardiac arrhythmias. Patients can be evaluated in the general paediatric clinic in the first instance with a thorough history and examination.

If there is good history for vasovagal syncope and the 12 lead ECG is normal, usually there is no need for further investigation and simple reassurance is all that is required. Increased salt and fluid intake and advice on posture when prodromal symptoms are experienced whilst standing can be helpful. Low dose fludrocortisone can be tried in some cases.

	• exertional syncope • associated chest pain on exercise • abnormal ECG • with history of previous out of hospital cardiac arrest • FH channelopathy • FH SCD/ PPM or ICD < 40yrs • triggered by fright/ surprise/ emotional stress • whilst swimming or drowning/near drowning • associated with *pathological* murmur	Cardiology review as inpatient where possible if patient is admitted *Call to discuss with on call team Aim for review in next 24 – 48 hours ETT at first review where possible
	• unexplained post exertional syncope • syncope whilst supine • abnormal ECG • associated with tonic-clonic or abnormal movements • FH of a genetic condition associated with sudden death e.g. LQTS or HCM • FH cardiomyopathy	Patient to be reviewed in cardiology outpatient clinic in next 8 weeks
	• Unexplained pre -syncopal symptoms	Patients are reviewed within the national 18-week framework by PEC or cardiologist

	○ history consistent with vasovagal syncope ○ Syncope or pre-syncope with a known non-cardiovascular cause ○ History consistent with reflex anoxic seizures	

Chest Pain

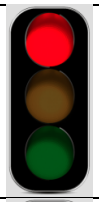
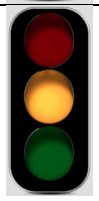
Chest pain is a common symptom in childhood and very rarely due to cardiac pathology. The vast majority are sharp musculoskeletal pains and can be evaluated in a GP surgery or general paediatric clinic and don't require further investigation.

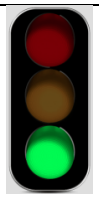
Referrals should focus on patient history (rarely are there clinical findings) and document:

- exertional or precipitating factors;
- relevant family history (FH).

Palpation of the chest (or deep inspiration) is useful to confirm a diagnosis of musculoskeletal pain that doesn't require further investigation or ongoing referral. Gastrointestinal disorders such as reflux or respiratory conditions may cause chest pain in children.

Inpatient referrals from a hospital setting should include a **resting ECG** where possible. In general, sending cardiac enzymes in children is not useful.

	Exertional chest pain with associated; ○ syncope ○ abnormal ECG Chest pain with recent illicit drug use	Cardiology review as inpatient where possible if patient is admitted *Call to discuss with on call team Aim for review in next 24 – 48 hours ETT at first review where possible
	○ associated murmur ○ exertional chest pain ○ other symptoms or signs of cardiovascular disease ○ non-exertional chest pain with abn ECG ○ FH of sudden unexplained death or cardiomyopathy ○ associated recent onset of fever	Patient to be reviewed in outpatient clinic in next 8 weeks

	• presumptively innocent murmur with no symptoms, signs, or findings of cardiovascular disease and a benign family history, normal ECG • family history of premature coronary artery disease	Patients are reviewed within the national 18-week framework by PEC or cardiologist
	• No other symptoms or signs of cardiovascular disease, a benign family history, and a normal ECG • Non-exertional chest pain with no recent ECG • Reproducible chest pain with palpation or deep inspiration	

Screening for Cardiac Disease Associated with Other Conditions

Depending on local provision, children who require ongoing cardiac surveillance may be seen in PEC, cardiology or cardiac physiology (echocardiography) clinics. **It is advised that all children at first assessment have a full echocardiogram to exclude associated CHD**. Ongoing surveillance and echocardiography may then be 'focused', or protocol driven, for example right heart assessment for pulmonary hypertension or left heart in patients with systemic hypertension.

Please note a number of tertiary specialised joint clinics run across the network and should be referred to where appropriate, for example metabolic cardiac clinic at RMCH.

1. Genetic

Patients with the following more common genetic diagnosis warrant a review in the paediatric cardiology clinic for assessment and echocardiography:

- Trisomy 21 (Down Syndrome) – these patients warrant assessment for the first 2 years to screen for evidence of pulmonary hypertension (even in the absence if CHD);
- 22q11 deletion (DiGeorge Syndrome);
- Trisomy 18 (Edwards Syndrome);
- Trisomy 13 (Pataus Syndrome);
- Noonans syndrome;
- Williams syndrome;

- Turners syndrome;
- CHARGE syndrome;
- VACTERL association.

The list is by no means comprehensive and covers the more commonly seen genetic conditions. For rarer conditions referral will typically be directed to cardiology by the clinical genetic service.

2. Congenital Anomaly

Many extra-cardiac anomalies can be associated with CHD. It is recommended that patients with the following conditions have a full echocardiogram. Many of these assessments will be done in neonatal and inpatient settings:

- Congenital diaphragmatic hernia (CDH)
- Exomphalos
- Gastroschiasis
- Ano-rectal malformation
- Tracheo-oesphageal fistula / oesophageal atresia
- Laterality disturbance (isomerism)
- Vein of Galen malformation

3. Neuromuscular and Neurology

Patients with many different neuromuscular conditions require intermittent cardiac evaluation from initial diagnosis, this includes:

- Duchenne muscular dystrophy (DMD)
- Myotonic dystrophy
- Friedreich's ataxia

Other conditions may present with more subtle cardiac dysfunction and cardiac evaluation is required from 12 years:

- Becker's muscular dystrophy
- Female carriers of DMD gene

Follow-up will generally be managed in consultation with the regional neuro-muscular team.

Patients with ischaemic stoke should be assessed with ECG and echocardiogram (to identify structural lesions and R to L shunts), depending on clinical presentation often a bubble contrast study (+/- Valsalva manoeuvre) is recommended.

4. Connective Tissue Disorders

Patients with suspected Marfan syndrome should be initially reviewed in the general paediatric clinic. Patients with the following diagnosis require ongoing cardiac review to screen for aortopathy and valvar dysfunction:

- Marfan syndrome
- Loeys Dietz
- Classical, cardiac-valvular, and vascular Ehlers Danlos Syndrome (EDS)

Other forms of connective tissue are not found to be associated with significant cardiac pathology and don't require referral:

- Benign Hypermobility Syndrome (EDS III)

5. Infective and Autoimmune

The following patients require referral to the paediatric cardiology clinic for assessment:

- Kawasaki Disease (see separate network guidance)
- Rheumatic Fever/Sydenham's Chorea

6. Respiratory Disease

Pulmonary hypertension may be seen in some children with respiratory disease. A selected group may warrant echocardiographic assessment:

- Right ventricular dominant pattern or right ventricular hypertrophy noted on resting ECG in respiratory patient;
- Severe chronic lung disease;
- Significant interstitial lung disease;
- Severe obstructive sleep apnoea.

7. Oncology

As part of their treatment regime a group of children will be treated with cardiotoxic chemotherapeutic agents. These treatment regimes and the echocardiographic surveillance required is clearly documented. Both AH and RMCH have streamlined pathways with cardiac physiologists providing screening echocardiography. It should be noted:

- A full echo assessment (documenting all the anatomy should be done at the start of treatment);
- Children with impaired function should be under review of a cardiologist in clinic.

8. Renal Disease

Patients with idiopathic systemic hypertension require a full cardiology assessment to look for evidence of CHD (particularly coarctation of the aorta).

Echocardiography may be advised if LVH on ECG, uncontrolled or undiagnosed hypertension or at transition onto adult services.

9. Endocrine Conditions

Patients with the following conditions under endocrinology follow-up require a full assessment at diagnosis and may require ongoing cardiac surveillance:

- Turners syndrome
- Congenital Hyperinsulinism (CHI)
- Hyperthyroidism

10. Haematology

In children with sickle cell disease echocardiography (to assess for pulmonary hypertension) should be arranged if there is evidence of chronic sickle lung disease, chronic unexplained hypoxia (oxygen saturations <95%) or other symptoms/signs suggestive of pulmonary hypertension.

11. Metabolic

Many paediatric metabolic conditions are associated with the development cardiac disease. This may present in the form of cardiomyopathy in conditions such as Pompe or Barth or with valvular disease in patients with Mucopolysaccharidoses (MPS). Referrals are typically made by metabolic medicine and optimal surveillance is in the joint clinic at Royal Manchester Children's Hospital.

Summary of Referral Pathway

Level 1: Specialist CHD Surgical Centre:
Alder Hey Children's Hospital

Level 2: Specialist CHD Cardiology Centre
Manchester Children's Hospital

Level 3: (PEC) Peripheral Cardiac Clinic run by Paediatrician/Neonatologist with expertise in cardiology

Level 3 Joint Clinic: (PEC and Cardiologist from Level 1 or 2)

Suspected Cardiac Anomaly picked up by GP, Neonatologist or Paediatrician

Depending on where the patient lives, refer to
(Care as close to home as possible)

Most common reasons for referral to Paediatric Cardiology:

- Murmur
- Cyanosis
- Chest Pain
- Palpitations
- Syncope or dizziness
- Family history of Congenital heart disease

Royal Manchester Children's Hospital
Specialist CHD Centre (Level 2)

Alder Hey Children's Hospital
Specialist Surgical CHD Centre (Level 1)

Local PEC Clinic
(Level 3)

F/u in PEC clinic

Discharge back to GP

F/u General Paediatrician

Refer General Paediatrician

Discharge Back to GP

Follow up in Level 1 or 2

Refer to Level 1 Specialist Surgical Centre
(Alder Hey Children's Hospital)

Follow up in PEC/Joint clinic

Discuss at MDT as required

Medical Management Regular F/u Level 1, 2 or 3

Surgery

Intervention

EP/Device

Second Opinion

Refer to Transplant/ PHT