Cardiology Advice and Referral Pathways – Paediatrics

A Clinical Guideline:

For Use In:	NICU, The Jenny Lind Department of Paediatrics, Blakeney ward, Children's Emergency Department		
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Version	Date	Author	Reason/Change
V6.0	09/03/21	Dr Rahul Roy and Dr Aravind Shastri	Document due for renewal
V7.0	01/05/2024	Dr Rahul Roy, Dr Aravind Shastri, Dr A Przystupa	Extended review- algorithms and more pathways added

Distribution Control

Printed copies of this document should be considered out of date. The most up to date version is available from the Trust Intranet.

Consultation

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 and recently in April 2023. additions regarding referral for Echocardiography and Innocent Murmurs. The new guideline has been discussed in the Paediatric Cardiology MDT meeting 25/01/24 and the circulated widely with paediatrics and neonatal teams and discussed in Governance meetings (NICU Governance May 2024 and Paediatrics Governance – June 2024)

Monitoring and Review of Procedural Document

The document owner is responsible for monitoring and reviewing the effectiveness of this Procedural Document. This review is continuous however as a minimum will be achieved at the point this procedural document requires a review e.g. changes in legislation, findings from incidents or document expiry.

Relationship of this document to other procedural documents

This document is a clinical guideline applicable to the Norfolk and Norwich University Hospitals NHS Foundation Trust.

Contents Page

Quick Reference Protocols: Incidental Murmur Quick Reference

Quick Referral for Echocardiography Quick Reference

PEG: Dr Aravind Shastri (AS)/Dr Edward Broad, PEC: Dr Rahul Roy (RR).

1. Introduction

1.1. 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 referrals for 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 appropriate 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 Paediatricians 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.

1.2. Objective

- To define the services offered by the local Paediatrician with expertise in Cardiology (PEC) and to provide guidance for common presentations.
- To define the services offered in the outreach paediatrician cardiology clinics run jointly with the tertiary centre Paediatric Cardiologist.

1.3. Scope

This guideline is for use by Paediatric Practitioners working at the Jenny Lind Children's hospital, in both the outpatient and inpatient settings, fo patients under 16 years of age.

1.4. Glossary

The following terms and abbreviations have been used within this document:

GD	Graham Derrick	SVT	Supraventricular Tachycardia
RR	Rahul Roy	QTc	Corrected QT interval
SB	Supriya Bhoomaiah	ICC	Inherited Cardiac Condition
AS/EB	Aravind Shastri/E Broad	WPW	Wolff-Parkinson-White
FM	Florian Moenkemeyer	ECG	Electrocardiogram
PEC	Paediatrician with Expertise	EEG	Electroencephalogram

	in Cardiology		
VSD	Ventricular septal Defect	MR	Mitral Regurgitation
ASD	Atrial Septal Defect	TR	Tricuspid Regurgitation
PS	Pulmonary stenosis	MVP	Mitral Valve Prolapse
AoS	Aortic Stenosis	GOSH	Great Ormond Street Hospital

2. Responsibilities

Dr Rahul Roy and Dr Aravind Shastri, Consultant Neonatologist and Paediatrician with expertise in cardiology, to update the guideline in line with current best practice. Paediatric Medical and Nursing Practitioners, to implement the following guidelines for appropriate referrals.

3. Clinical Guidelines

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).

Care Pathway- Identification to Diagnosis

Useful referral details

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

Out of hours - There is no dedicated on-call service but discussion with GOSH cardiology registrar is strongly encouraged in confirmed/strong suspicion of CHD/acquired heart disease.AS/EB (paediatrician with expertise in cardiology) or RR (Neonatal consultant with expertise in cardiology) can be contacted by Consultant on-call (directly or via switchboard and if they are available, an echocardiogram can be facilitated); Extra contractual hours can be recognised by renumeration or TOIL

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/EB/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.

3.1. Heart Murmurs and Congenital Heart Disease

3.1.1. Innocent Heart Murmur

Heart murmurs in Children are very common and are often identified during a routine assessment for an unrelated presentation. Murmurs in asymptomatic and otherwise healthy children are likely to be ‘innocent’, due to the normal flow of blood through the heart and vessels. It is unlikely major/complex or life-threatening conditions would present with an asymptomatic murmur after 6 weeks of age. Differentiating innocent murmurs from structural heart disease is largely clinical, ECGs and CXRs do not significantly contribute to the diagnosis.

Innocent Murmur Features:

- Asymptomatic
- Soft blowing murmur
- Systolic Murmur only, not diastolic.
- Left Sternal edge.
- Normal heart sounds, with no added sounds.
- No parasternal thrill.
- No radiation.

All areas must be auscultated: Mitral, Tricuspid, Pulmonary, Aortic and radiation areas.

Concerning Murmur Features:

- Diastolic murmur

- Loud systolic murmur, i.e. ≥Grade 3/6 with thrill, long in duration, transmits well to other parts of the chest.
- Abnormal heart sounds, systolic click or opening snap.
- Central cyanosis.
- Abnormal peripheral pulses.
- Continuous murmur that cannot be suppressed.
- Abnormal ECG.

The following flowchart may be used when an incidental murmur is found and the child is thriving, cardio-vascularly stable, has no features of heart failure, has normal pulses **(*including femorals) and is acyanotic:***

Innocent murmur leaflet – see Appendix 3

3.1.2. Small Mid-muscular & 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 peri-membranous 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 AS/RR clinic until it undergoes spontaneous closure. Peri-membranous VSD requires review with GD/FM.

3.1.3. 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.

3.1.4. Pulmonary Stenosis

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

Classification of stenosis:

Velocity:

- 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 6 -18 months.

3.1.5. Aortic Stenosis (AoS)

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

Classification of stenosis:

Velocity:

- 2-3 m/s – mild AS
- 3-4 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.

3.1.6. 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.

3.1.7. Mitral Valve Prolapse

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.

3.1.8. 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.

3.1.9. 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.

3.1.10. 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.

3.1.11. Family History of Congenital Heart Disease/Inherited Cardiac Conditions

Please Refer to table 1 and key messages at the end of this document.

3.2. Chronic Lung Disease of Prematurity

Patients needing nasal cannaula oxygen 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 the 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 O_2 requirement worsening after discharge, repeat Echo in 4 months; if persistent O_2 even at 1 year of age, another echocardiogram can be requested.
- Those needing HHFNC treatment at 36 weeks CGA or nasal cannula O_2 (> 0.5L/min to 1L/min) even at 36 weeks CGA will need earlier Echo (between 34-36 weeks).

3.3. 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.

Cardiac type of Chest pain can be associated with structural lesions of the heart, in particular aortic stenosis, aortic dissection (Marfan's syndrome) and mitral valve prolapse or very rarely coronary artery abnormalities. 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.

Please see chest pain algorithm below on pages 21-23

3.4. Palpitations

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-excitation 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.

3.5. 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.

3.6. Inherited Cardiac Conditions

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.

3.7. Referral for Echocardiography

Please use the algorithm below when referring patients for Echocardiography. Should the PEC not be available, please liaise with the GOSH Cardiology Team regarding necessity of the Echo and potential for transfer to GOSH for further investigation.

In exceptional circumstances, AS/RR/SB may be available Out of Hours/Weekends for Echocardiography, but this would require a Consultant-to-Consultant referral.

PEG: Dr Aravind Shastri (AS). PEC: Dr Rahul Roy (RR).

3.8. 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.

Guidance for Paediatricians to Triage or Refer patients to Paediatrician Cardiology clinics in the Outpatient Referral Console

Cardiac Murmurs – suggestive of a cardiac cause

- **Pathological murmurs** – loud i.e., Systolic Grade 3/6 & any Diastolic murmurs.
- **Murmur associated** with:

  Central Cyanosis.

  Failure Symptoms – Tachypnoea, Tachycardia, FTT, poor feeding & weight gain, increased sweating, Frequent respiratory infections,

  Abnormal peripheral pulses.

  NOTE: Functional murmurs are caused by extracardiac diseases that results in a change in blood flow phenomena as a result of fever, severe anaemia – **Gen Paediatric Clinic Review.**

Chest Pain – warning signs for an underlying cardiac disease.

- Increases with exertion.
- Associated with syncope and/or palpitations suggestive of cardiac cause
- Positive f/h of Cardiomyopathy, Marfan Syndrome, long QT syndrome, sudden deaths in **first degree relative.**

Palpitation – suggestive of cardiac cause.

- Sudden onset and abrupt end are a sign of paroxysmal SVT.
- Associated with syncope, chest pain.
- Occurring when the child stands up suddenly or after standing for a long period – POTS.

Syncope – suggestive of a cardiac cause.

- During physical exertion.

- Without prodrome.
- After a loud noise or unexpected sound.
- In cold water/whilst swimming.
- Whilst lying down.
- Prolonged >5min.
- Positive f/h of Sudden Cardiac Death (SCD) in first degree relative (Parents, siblings & direct offspring's).

Vasovagal Syncope – Paediatrician clinic follow up.
NOTE: **Vasovagal syncope** is not an indication for cardiology clinic referral.

Suspected Inherited Cardiac Conditions – (Cardiomyopathy, Aortopathy, LQTS, Brugada, Marfan Syndrome, Bicupsid Aortic Valve)

- Positive family history in **FIRST degree relatives**

Abnormal ECG associated with cardiac signs & symptoms – Please be cautious with automated ECG interpretation in Paediatric patients.

3.9. Echocardiography for children with Genetic Syndromes

Echocardiography schedule for Children with suspected/diagnosed Genetic Syndromes in the presence of a normal cardiac examination.

Down's Syndrome (DS)

- Echocardiogram and ECG before discharge home.
- Paediatric Cardiology review and Echocardiography within 6-8 weeks of birth, 1 year and 2 years of age even in patients without CHD.
- Auscultation of the heart annually after 2-years.
- Echocardiogram for new murmurs and signs of cardiac failure.
- A single Echocardiogram in adult life.
- *The cardiac problems with DS are characterised by Pulmonary Hypertension and Biventricular Dysfunction. Pulmonary Hypertension is prevalent in children with DS with/without CHD. Infants with DS have persistent, abnormal elevation of PVR over the first 2 years of age, regardless of the presence or absence of structural congenital heart disease.*

The Heart - Downs Syndrome Association (downs-syndrome.org.uk)

Turner's Syndrome (TS)

- Echocardiography at diagnosis.
- Common associations – Bicuspid aortic valve, Coarctation and Aortic dilatation.

- If none of the above → 5-yearly Echocardiography in Children and 10-yearly in Adults.
- For Turner's Syndrome with no structural heart disease annual assessment of Blood Pressure. ECG should be completed to assess for potential conduction and repolarisation abnormalities.

Noonan's Syndrome (NS)

- Echocardiogram at diagnosis.
- Annual Echo until the age of 3 years. Then at 5 years, 10 years and adolescence (due to risk of Cardiomyopathy).
- Ongoing Cardiology follow-up required into adulthood.

NS Guidelines 7 FINAL (rasopathiesnet.org)

William's Syndrome (WS)

- At diagnosis for neonates.
- Paediatric Cardiology Echocardiography must be completed within 3 months for infants.
- Annual cardiac examination by a paediatric cardiologist should be carried out until 4 years of age.
- Thereafter, complete cardiac assessment including Echocardiography at least every 5 years.

DRAFT 18 WSF (williams-syndrome.org.uk)

DiGeorge Syndrome
Echocardiography at diagnosis and if normal, no further follow up necessary.

3.10. Maternal SLE (anti-Ro and anti-La antibodies)

Neonatal Lupus is a passively acquired autoimmune disease, caused by passage of maternal anti-Ro/SSA and/or anti-La/SSB antibodies across the placenta.

Signs of Neonatal Lupus include:

- A red, raised rash on the scalp and around the eyes. This typically resolves by 6-8 months of age as the maternal antibodies are cleared.
- Transient abnormalities of liver function.
- Generalised or selective reduction in cellular blood components. E.g Anaemia, thrombocytopenia.
- Complete heart block. Rarely associated with Cardiomyopathy.
- (Heart block may be noted antenatally).

Management of the asymptomatic Baby born to Mothers with anti-R/La

- Clinical Examination soon after birth looking for the clinical features of lupus and assessing the heart rate. If the rate is >80 beats per minute and the Baby is clinically stable, they should remain with Mother.
- Full Blood Count and differential WBC.
- ECG prior to discharge. This should be discussed with the Neonatal Consultant prior to discharge.
- Encourage Breast-Feeding. There is no increased risk of Neonatal Lupus with Breastfeeding.

- Follow-up: Out-patient clinic in 2-3 weeks. The family should be advised to contact the Neonatal Unit earlier if any rash develops or there are other parental concerns.
- Advise to avoid Baby exposure to direct sunlight.

Mother's on Biologics will need discussion with Neonatal Consultant.

Management of the symptomatic Baby born to Mothers with anti-R/La

Non-cardiac manifestations of Neonatal Lupus are generally transient and resolve with the clearance of maternal antibodies. However, damage to the conduction system and myocardial disease are irreversible.

Babies with a slow heart rate (<80BPM) should be transferred to the Neonatal unit for baseline ECG and a 24 hour tape.

The Baby should be discussed with the on-call tertiary Cardiology team.
If there are signs of low cardiac output, isoprenaline may be useful to temporarily increase the heart rate, prior to pacemaker insertion.

Pancytopenia typically resolves over 4-6 months, however, IV Immunoglobulin may be helpful in certain cases. Discuss FBC abnormalities with the Neontal Consultant.

Maternal SLE advice taken from the Greater Glasgow and Clyde guidelines. Please follow the following link for further details: Anti-Ro & Anti-La antibodies : Guideline for the management of babies born to mothers with systemic lupus erythematosus (SLE) and other autoimmune disorders (scot.nhs.uk)

4. Related Documents

Heart Murmurs in Newborn Babies (Trustdocs ID: 1223).

Trust Guideline for Referral when a Fetal Abnormality is detected" – Trustdocs ID: 882 (guideline number AO16).

5. References

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6. Monitoring Compliance

Compliance with the process will be monitored through the following:

Key elements	Process for Monitoring	By Whom (Individual / group /committee)	Responsible Governance Committee /dept	Frequency of monitoring
Referral to the Paediatrician cardiology clinic (RR/AS/EB) should strictly comply with the cardiology referral pathway**	Audits Review of referrals and echocardiography data from cardiac physiologists	Paeds services manager/ Admin staff/ Dr Shastri/Dr Roy	Paediatric governance	3-5 yearly
** At the time of writing this review cardiac physiologist led Echocardiography service is not running on weekdays and for urgent needs; is being run by a locum physiologist on weekends only; hence there are increasing referrals to paediatric consultant led clinics and thus not able to strictly follow referral pathways.				

The audit results are to be discussed at the Paediatric governance meetings review the results and recommendations for further action. Then sent to trust audit committee/Paediatric directorate who will ensure that the actions and recommendations are suitable and sufficient.

7. Appendices

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 Also request Neonatal Assessment Clinic for this date. The ECG should be reviewed for acute pathology and delivered to the named Consultant. Refer only if abnormal.

Heart murmurs in family***	No	No	As standard	No

Appendix 1 - 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 a first degree relative **needing cardiac surgery/intervention,** the pregnant lady will be referred to the 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/Coarctation 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 and 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 (E.g. 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.

Appendix 2 – Chest pain children (guidance for ED and Paediatric teams)

Rationale

Chest pain is a common presenting complaint in children and young adults. As the large majority are benign in nature, significant pathology can be overlooked, it is therefore prudent to have a generic guideline in place to support clinicians during

their assessment. It is important to note that this guideline should be used solely as an aide and if clinician concern persists, this should be acted upon appropriately.

Background

Chest pain is a common presenting complaint in children and young adults. The large majority of these patients do not have either cardiac (< 1%) or other serious pathology. The aim of initial assessment should be to exclude life-threatening pathology and subsequently form a diagnosis and management plan for the remaining cases.

Common causes of chest pain in low risk patients include:

- Musculoskeletal strains/costochondritis
- Respiratory infections
- Asthma exacerbations
- Gastritis/reflux/biliary disease
- Precordial catch syndrome – sudden, sharp + short-lived chest pains in young healthy teenagers
- Anxiety
- Idiopathic – many well children will have no organic diagnosis made

Risk Factors for serious pathology (+ potential pathology)

- First episode of pain
- Previous cardiac disease or surgery
  - *Myocardial ischaemia, arrhythmia, pericarditis, pericardial effusion*
- Hypercoagulable states: primary clotting disorder, neoplasm, pregnancy, oral contraceptive, central lines, post-operative
  - *Pulmonary embolus*
- First degree family history of cardiac disease: early/sudden cardiac deaths of unknown cause, arrhythmias, cardiomyopathy
- Sickle cell disease
  - *Acute chest syndrome*
- Chronic respiratory disease
  - *Pneumothorax*
- Kawasaki Disease (previous or current)
  - *Coronary aneurysm + myocardial ischaemia*
- Familial hyperlipidaemia
  - *Myocardial ischaemia*
- Cocaine or stimulant use
  - *Myocardial ischaemia*
- Connective tissue disorders: Marfan Syndrome, Ehlers Danlos Syndrome
  - *Pericarditis, pericardial effusion, aortic dissection*
- Other genetic disorders: Turner Syndrome, Down Syndrome

Chest pain Assessment

Any acutely unwell patient should be assessed as per APLS guidelines using the ABCDE approach.

If a clear diagnosis e.g. costochondritis is indicated from the history and examination, routine investigations are not necessary. Children without a clear cause for their chest pain or who have risk factors should have a 12 lead ECG. Chest X-ray should be done selectively when a specific pathology is suspected e.g. empyema or pneumothorax. Chest X-ray should not be used as routine screening without a specific indication

Features prompting further investigation from the history include palpitations or collapse.
Concerning features on examination include – respiratory distress, abnormal vital signs including tachycardia, poor perfusion, distended neck veins, muffled heart sounds + altered mental state.

The below flow chart provides a suggested guide for assessment, if any concerns that a patient is unwell or high risk then discuss with a senior clinician.

Referrals + Onward Management for specific cases

1. Undifferentiated chest pain +/- benign palpitations + normal ECG – GP follow-up, no need for Paediatric referral
2. History + ECG suggestive of Pericarditis (appendix 1)
    a. Persistent pain despite NSAIDs – admit to CAU for observation, give NSAID + echo if available.
    b. Pain settles with analgesia - can be discharged with regular NSAID for 7 days, urgent Cardiac follow-up (internal referral) and open access until appointment.
3. History + ECG (+ Troponin) suggestive of Myocardial Ischaemia – admit to HDU, urgently discuss with tertiary Cardiology (GOSH) and consider transfer. Manage as per MI; Morphine, Oxygen, Aspirin may be indicated – commence only after discussion.
4. History + examination suggestive of Marfan's (modified Ghent criteria – score > 5, appendix 2)
    a. Ongoing severe chest pain and abnormal PEWS consider CTA-Aorta following discussion with ED/CAU Consultant.
    b. If pain persistent with normal PEWS – admit with continuous observations.
    c. If pain resolved and normal PEWS – refer to Cardiology clinic (via internal referral)

Pediatric Chest Pain – Quick Reference Guide

Appendix 3 – Innocent heart murmur patient information leaflet

This leaflet gives you general information about innocent heart murmur. It does not replace the need for personal advice from a healthcare professional. Please ask us if you have any questions.

What is an innocent heart murmur?

When you listen to the heart through a stethoscope, you can hear the sounds of the heart working – blood pumping through the chambers of the heart and the heart valves opening and closing. Normally, these sounds form a clear, regular pattern. A heart murmur, on the other hand, is an unusual sound, which can be heard when listening to the heart. Heart murmurs are very common. They can be a sign of a heart problem, but your child's murmur has been diagnosed as "innocent".

This means that the murmur is harmless and is not a sign of a heart problem. Sometimes innocent heart murmurs are called as below.

Vibratory murmurs, Venous hums, Functional murmurs

What causes innocent heart murmurs?

When we are young, our hearts are quite small and compact. This means that the bends in the heart around which the blood has to flow are quite tight. To push blood around these tight bends, the heart has to beat quite quickly – much faster than it does in adults. An innocent heart murmur is usually the sound of the blood being pushed around these tight turns. It is similar to the noise a stream makes as water curves around a bend. Innocent heart murmurs are louder when the heart is beating faster than usual. For example, the heart can beat faster if your child is anxious or has a high temperature.

Will my child need any treatment?

As your child's heart murmur is innocent, there is no need for any further visits to the cardiologist or for any special care or treatment. Most innocent heart murmurs eventually disappear as a child grows older and his/her heart grows bigger.

Summary

- Innocent murmurs can happen in healthy hearts
- No special precautions are needed
- No follow-up treatment is needed

Equality Impact Assessment (EIA)

Cardiology Advice and Referral Pathways – Paediatrics

Type of function or policy	Existing

Division	Women and Children	**Department**	Paediatrics
Name of person completing form	Dr Aravind Shastri	**Date**	08/07/2024

Equality Area	**Potential Negative Impact**	**Impact Positive Impact**	**Which groups are affected**	**Full Impact Assessment Required YES/NO**
Race				No
Pregnancy & Maternity				No
Disability				No
Religion and beliefs				No
Sex				No
Gender reassignment				No
Sexual Orientation				No
Age				No
Marriage & Civil Partnership				No
EDS2 – How does this change impact the Equality and Diversity Strategic plan (contact HR or see EDS2 plan)?	Does not affect			

- **A full assessment will only be required if: The impact is potentially discriminatory under the general equality duty**
- **Any groups of patients/staff/visitors or communities could be potentially disadvantaged by the policy or function/service**
- **The policy or function/service is assessed to be of high significance**

IF IN DOUBT A FULL IMPACT ASSESSMENT FORM IS REQUIRED

The review of the existing policy re-affirms the rights of all groups and clarifies the individual, managerial and organisational responsibilities in line with statutory and best practice guidance.