

Screening for Congenital Heart Disease (CHD) Screening - Full Clinical Guideline

Reference no.: NIC SS 12/July23/V8

1. Introduction

The incidence of congenital heart disease (CHD) is 8/1000 (0.8%) births, with 20-30% in the UK considered critical, i.e requiring surgery within the first year of life. The UK has 14 NHS population screening programmes including the Fetal Anomaly Screening Programme (FASP) which may identify ~51% of critical congenital heart conditions, and the NHS Newborn and Infant Physical Examination (NIPE)¹. The NIPE allows for prompt identification of potentially life-threatening lesions and should be completed by 72hrs of life in babies born after 34 weeks, or as soon as possible after reaching 34 weeks, if no contra-indications. NIPE is performed by trained health practitioners or doctors. It is repeated by the General Practitioner 6-8weeks later.

Detection of CHD in the newborn may be influenced by their transitioning from in-utero to ex-utero life; a NIPE done, or baby discharged before 24hrs of age may not have completed its transition and parents counselled accordingly.

2. Aim and Purpose

This guideline summarizes UHDB's pathway for detection, assessment, and management of CHD on our postnatal wards, and transitional care if applicable). It does not cover specific CHD lesions or babies admitted to the neonatal unit, children's ward or discharged home who re-present with suspected CHD. It incorporates Neonatal Heart Murmurs WC/PN/30N, version 7 which has been archived.

3. Definitions, Keywords

AVSD:	Atrio-ventricular septal defect
ACP:	Advanced Clinical Practitioner
CHD:	Congenital Heart Disease
CXR:	Chest x-ray
ECG:	Electrocardiogram
EMCHC:	East Midlands Congenital Heart Centre
PEC:	Paediatrician with Expertise in Cardiology
PNW:	Postnatal ward
NEWTT:	Newborn Early Warning Trigger and Track
NICU:	Neonatal Intensive Care Unit
NIPE:	Newborn and Infant Physical Examination
UHDB:	University Hospitals of Derby and Burton

4. Main body of Guidelines

1) **The Guideline:** For ease, we have divided the guideline into five (5) groups.

Group 1:

These are babies **identified on antenatal scan as having significant CHD**, having had detailed specialist fetal cardiology assessment and opinion. These cases are discussed at the Joint neonatal/ Paediatric feto-maternal liaison meetings, with individualized plans agreed with a copy filed in a dedicated "Expected Babies" file on NICU in Derby and neonatal alert form in Burton.

Group 1 action:

- i. When the baby is born, follow individualized plan as documented above.
- ii. The baby will require urgent medical review within **30mins** of birth, and management as per plan.

Group 2:

NIPE with abnormal symptoms and or signs, may sometimes be due to CHD; the symptoms include central cyanosis, tachypnoea/respiratory distress, apnoeas >20s, absent/weak femoral pulses, heaves/thrills/visible pulsations or significant murmurs (usually loud, harsh and widely heard).

Group 2 action: This group needs rapid evaluation and should not be discharged home, without a clear diagnosis and management plan.

A) All symptomatic (NIPE positive) babies should be assessed for cardiac or non-cardiac causes and treated as clinically indicated.

- i) Do pre- and post-ductal saturation before discharge, **See section 6 A) below.**

B) Further OPD review depends on if a CHD is still suspected or confirmed.

Group 3:

Some babies on NIPE may have dysmorphic features associated with higher risk of CHDs; they may have no symptoms. Action will depend on the presence or absence of a murmur.

Group 3 action:

C) with a murmur:

- i) they should be reviewed promptly by the tier 1 neonatal/Paediatric team and
- ii) Inform the senior colleague as soon as possible, and or consultant if appropriate.
- iii) do pre- and post-ductal saturation as part of the assessment, **See section 6 A) below.**
- iv) **Follow-up** depends on the clinical findings or consultant decision.

D) With no murmur:

- i) observed for 24hrs on the postnatal ward with NEWTT as protocol.
- ii) a tier 1 review before discharge home
- iii) do pre- and post-ductal saturation before discharge, **See section 6 A) below.**
- iv) Arrange Paediatric cardiology OPD or discuss with senior before discharge.

Group 4:

Asymptomatic murmurs on NIPE, well babies with no risk factors for CHD.

Group 4 action: they require evaluation prior to discharge. The guiding principles are:

- i) Baby with a murmur should be at least 24hrs old before discharge.
- ii) Start NEWTT when a murmur is heard and stop on review by the neonatal team if no other concerns.
- iii) A baby with a murmur but no other clinical concerns should be reviewed after 24hrs of age but within 24hrs of hearing the murmur.

The **suggested actions** thus are:

- iv) If a murmur is heard and baby is less than 24hrs old, we recommend the baby is not discharged until they are at least 24hrs old. If parents insist on going home against medical advice, a pre/post ductal saturation should be done, and baby may be discharged if they are normal. If an abnormal reading, the baby should not be discharged even against medical advice; in this instance the Neonatal registrar and Midwifery Manager on call should be informed to see the baby and their parent (s).
- v) On hearing a murmur NEWTT obs should commence until the baby is reviewed by the neonatal team and at least 24hrs old.
- vi) The neonatal team should review the baby within 24hrs of the murmur but does not need to wait a further 24hrs from when the murmur was heard before the review; however, the baby needs to be at least 24hrs old.
- vii) If on review the murmur has resolved, the NEWTT obs initiated because of the murmur should stop if the baby is over 24hrs old.
- viii) If the baby is over 24hrs old and murmur remains, NEWTT can stop if the baby has normal pre/post-ductal saturation and there are no other concerns. The baby can be discharged home with review in the murmur clinic within 4 weeks.
- ix) If the baby becomes **symptomatic**, request urgent neonatal/paediatric review.

Group 5:

Asymptomatic with family History of CHD: The recurrence risk for siblings of children with any CHD is estimated at 1–4%, higher if strong family history of a particular malformation. A history of congenital heart disease in a first degree relative should result in an antenatal referral to the Fetal Medicine department for a detailed scan rather than a routine antenatal screening. A family history of congenital heart disease is recorded as a risk factor in NIPE SMART.

- i) Family history in first degree relative of **Left Ventricular Outflow Obstruction** (Hypoplastic left heart, coarctation aorta or bicuspid aortic valve)
- ii) Family History of **Congenital Cyanotic Cardiac defects** in first degree relatives
- iii) Family history of **cardiomyopathy** in first degree relatives
- iv) Family history of **Brugada, Long QT** in first degree relatives.
- v) Family history of surgically corrected ASD or VSD, in first degree relatives

Group 5 action:

- i) If family history of structural heart defects, perform and document on the referral a pre and post ductal saturation.
- ii) Refer to Paediatric cardiology, asymptomatic babies with above family history.
- iii) Babies with first degree relatives with "hole" in the heart, Wolfe Parkinson White (WPW), Atrial fibrillation should not be referred. A pre-discharge 12 Lead ECG is not usually helpful. If unsure of the family history, advice parents of asymptomatic babies, to discuss with their GP.

2) Non urgent pre-discharge paediatric cardiac review

Prior to discharge, though not limited to, it is desirable to have a senior paediatrician/ACP or Paediatric cardiology review in these group;

- A) confirmed or suspected trisomy 21
- B) as advised on the neonatal alert
- C) as clinically agreed by the admitting consultant and local PEC or paediatric cardiologist

3) Urgent paediatric cardiology review, either in house or via tertiary paediatric cardiology center

- A) As advised on the neonatal alert
- B) Baby with saturation < 96% despite FiO2
- C) Baby with absent femoral pulse
- D) As clinically indicated after review by the admitting consultant and agreed with a PEC or EMCHC. Arrange transfer of baby to Cardiac Centre for further evaluation if unable obtain one locally. Inability to access a UHDB PEC should not delay seeking help from the cardiac center.

4) Parent information leaflet: Parents of any baby sent home with a murmur, should have written parent information leaflet on murmur, explaining the "Red flags" to look out for and how to seek medical advice prior to the OPD appointment, given to them.

5) Relevant Investigations

- A) **Oxygen saturation:** for the purpose of screening, a level <96% on air or a difference of >3% between the pre and post ductal saturations is **failed** screen.
 - i) All failed screen, must get urgent senior review and or admitted to the neonatal unit
- B) **12 lead ECG:** not typically informative.
 - i) May be useful in suspected asymptomatic Trisomy 21 if no access to echocardiogram prior to discharge. It may show a superior axis of AVSD.
 - ii) Do pre-Discharge ECG if abnormal pulse rate noted during labour or NIPE.

- C) **CXR:** only if clinically indicated.
- D) **4 limb BP:** may be useful especially in symptomatic babies or suspected coarctation. A normal 4 limb BP does not exclude coarctation, nor does abnormally raised confirm coarctation.
- E) **Echocardiography:** useful tool in the hands of appropriately trained operator.

6) **Obtaining in-house echocardiography**

Central to the success of this screening pathway is the availability of **Echocardiography** and access to specialist cardiac assessment and management by the UHDB PEC team. This should be a consultant-to-consultant request, and if available and necessary, a PEC will perform an echocardiogram, prior to discharge from postnatal ward. If a pre-discharge paediatric cardiology assessment is not possible, and baby is well, inform the admitting neonatal or paediatric consultant, and make a detailed referral to Paediatric cardiology team for the next available cardiac clinic.

- a) **Queens Hospital Burton:** via the relevant secretary for paediatric cardiology.
- b) **Royal Derby Hospital:** Postnatal ward referral form or Neonatal ward clerk

7) **Suggested audit standards.**

Recommend regular cross site audit of this guideline, at least once every 3 years prior to renewal or following a significant interim review.

- i) **100% of group 1** babies should have antenatal plan accessible prior to birth
- ii) **100% of group 1** babies should be evaluated within 30 minutes of birth.
- iii) **100% of babies** with suspected CHD kept on the PNW for a further 24hrs must have regular NEWS.
- iv) **100% of babies** discharged home with a murmur must have a normal pre- and post-ductal saturations prior to discharge. i. e<3% difference and >95% on air and documented on the referral.
- v) **100% of babies** discharged home with a murmur should be seen in clinic by 4 weeks old.

5. **References**

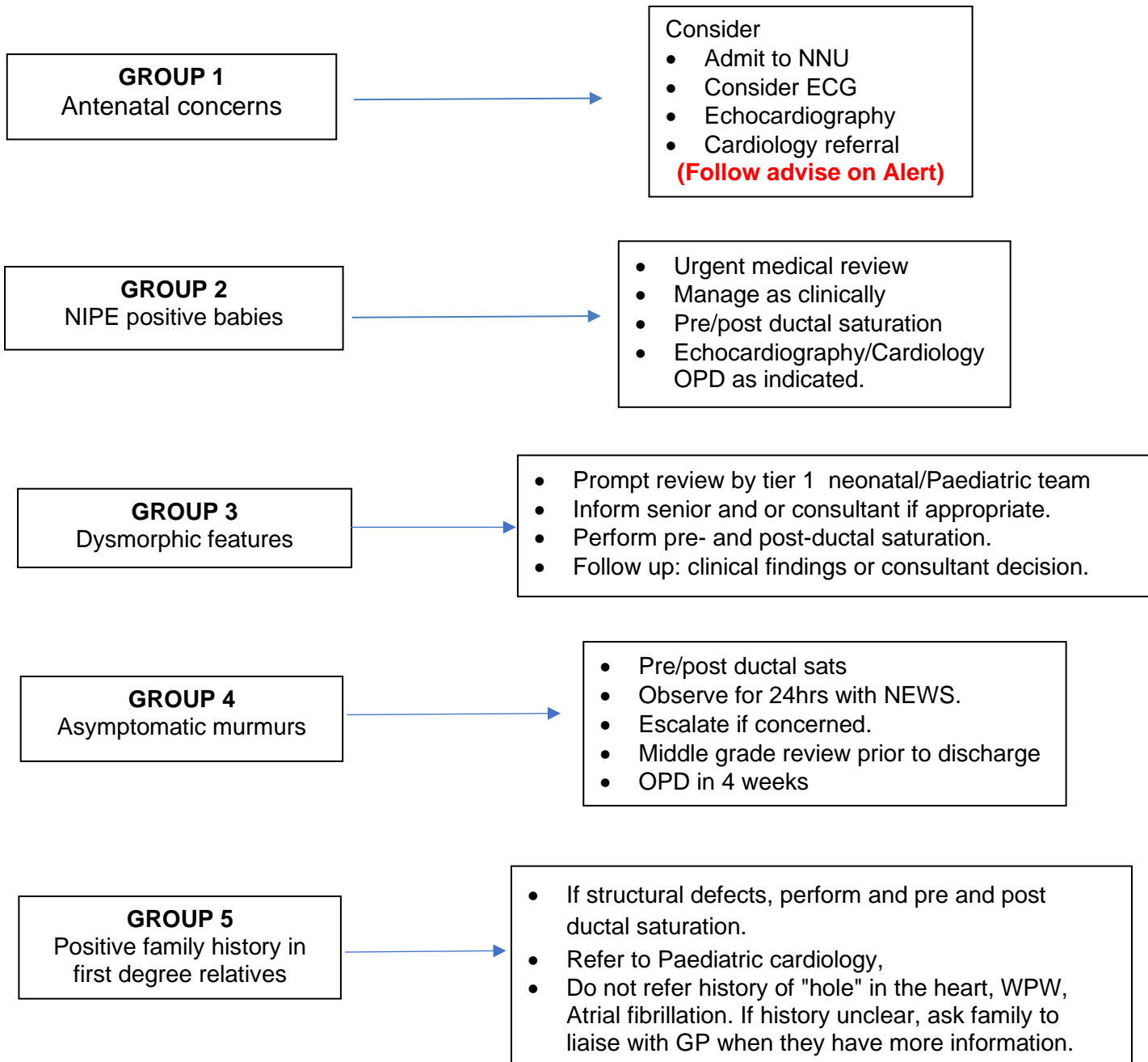
Newborn & Infant Physical Examination – Screening Programme Standards 2021

6. Documentation Controls

Reference Number NIC SS 12	Version: V8		Status Final	
Version / Amendment History	Version	Date	Author	Reason
	Neonatal Heart Murmurs WC/PN/30N, version 7	06/07/2023	Dr B Etuwewe	1. Adopt single trust wide guideline 2. Out of date
	V8	25/07/2023	Dr B Etuwewe	3. Minor amendments to guideline
Intended Recipients: All Paediatric, Neonatal, Midwifery or obstetric staff responsible for assessing and screening the newborn for possible heart conditions.				
Training and Dissemination: Via governance structures to all UHDB Paediatric, Neonatal, Midwifery or obstetric staff responsible for assessing and screening the newborn for possible heart conditions.				
Development of Guideline: Dr Bemigho ETUWEWE Job Title: Consultant in Neonatology and General Pediatrics (with Expertise in Paediatric Cardiology)				
In Consultation with: Neonatal team (RDH) Paediatric team (QHB) NIPE Midwives (QHB/RDH) Consultant Obstetrician/foetal medicine (QHB/RDH)				
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7. Appendices

a. SUMMARY OF CHD SCREENING



b. Neonatal Heart Murmurs

