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Disclaimer:

This clinical Guideline was produced after careful consideration of available evidence in conjunction with clinical expertise and experience. It is intended that Trusts within the region will adopt this guideline and educational resource after review through their own clinical governance structures. This guideline does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient.

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Prenatal Diagnosis of Congenital Heart Disease and Perinatal Management Planning SOP

4th June 2024



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Introduction

Congenital Heart Disease (CHD) may be identified prenatally with a high level of diagnostic accuracy in the right circumstances. For some CHD conditions there is evidence that improved pre-operative condition translates into better postnatal outcomes. Furthermore, early diagnosis during pregnancy will allow parents to consider their options and make decisions about whether to continue with the pregnancy. For parents continuing with pregnancy, they can gather information about the child's condition and be prepared for subsequent surgery and treatments. Most cases of CHD occur in low-risk pregnancies and will only be detected by screening at the time of obstetric ultrasound scans.

There are two sets of standards that relate to the screening and diagnosis of CHD. These are the Fetal Anomaly Screening Programme (FASP) and the BCCA Fetal Cardiology Standards. FASP sets out the standards for the initial anomaly ultrasound scan that may identify a possible congenital cardiac abnormality. The BCCA document details the standards for the fetal cardiology assessment. The BCCA and the FASP standards overlap at the point of entry into the fetal cardiology pathway and therefore the two are not mutually exclusive of each other, rather the two pathways are continuous and may be used together.

Fetal Anomaly Screening Programme (FASP)

NHS FASP, based in Public Health England, is an expert team that ensures national consistency and provides expertise.

NHS FASP requires that there is equal access to uniform and quality-assured screening across England and that women are provided with high quality information so they can make a personal informed choice about their screening and pregnancy options.

The FASP Screening programme happens at key points along pregnancy but essential to congenital heart disease is the anomaly scan performed at around 20 weeks gestation.

The anomaly scan is designed to identify anomalies which indicate:

- > Conditions that may benefit from treatment before or after birth
- > That the birth should occur in an appropriate hospital/centre
- > That the baby's treatment is optimised after the baby is born
- > Babies that may die shortly after birth.

Some women may choose not to be screened at all and it is important that this choice is respected.



Anomaly Scan

NHS FASP recommends the offer of a mid-pregnancy scan which is undertaken between 18⁺⁰ to 20⁺⁶ weeks of pregnancy to screen for major fetal anomalies.

Some providers choose to arrange the fetal anomaly scan later within the recommended window, that is closer to 20 weeks as opposed to 18 weeks. Where this occurs, services must be able to facilitate referrals for further investigations and options for pregnancy choices in a timely manner and within the required national timeframes. Ongoing audit of practice should be in place to monitor conformity. The screening pathway must be completed by 23⁺⁰ weeks of pregnancy.

Women who wish to have a fetal anomaly ultrasound scan, but do not wish to be informed if abnormalities are found, should be advised that all significant findings seen on the scan will be reported and therefore they should consider not having fetal anomaly ultrasound screening.

The main structures to be assessed at the 18⁺⁰ to 20⁺⁶ week scan are defined as shown in Table 1. Abnormalities of these structures can indicate a number of specific conditions. Other conditions may be detected using this ultrasound screening test, but there is insufficient data to confidently predict the standard which should be achieved. Currently FASP are monitoring the prenatal detection rate for the following cardiac lesions, their set thresholds for detection are below

- > transposition of the great arteries – acceptable $\geq 70.0\%$, achievable $\geq 99.0\%$
- > atrioventricular septal defect – acceptable $\geq 50.0\%$, achievable $\geq 80.0\%$
- > tetralogy of Fallot – acceptable $\geq 55.0\%$, achievable $\geq 85.0\%$
- > hypoplastic left heart syndrome – acceptable $\geq 80.0\%$, achievable $\geq 99.0\%$

Fetal Cardiac Protocol

During the ultrasound the following views are required:

1. Situs/Laterality
2. Four-Chamber: Transverse section of the thorax including a complete rib and crux of the heart
3. Aorta/Left Ventricular Outflow Tract: This view shows the outflow tract of the left ventricle
4. Pulmonary/Right Ventricular Outflow Tract: this view shows the outflow tract of the right ventricle only. The Three-Vessel View (3VV): this view shows the outflow tract of the right ventricle including the pulmonary artery and its relationship to the aorta and superior vena cava.
5. The 3 vessel and trachea view (3VT): a transverse view of the fetal upper mediastinum; it depicts the main pulmonary artery in direct communication with the ductus arteriosus, the transverse aortic arch and the superior vena cava, together with their relationship to the fetal trachea.

A single repeat scan must be offered and completed by 23+0 weeks gestation in cases where the image quality of the first examination is compromised. The repeat scan can be offered on the same day, or a new appointment arranged according to local assessment.

Where there are inadequate views of the cardiac components of the anomaly scan, FASP guidance recommends that the screening anomaly scan be repeated within 2 weeks. If the cardiac views still cannot be completed, then the limitation of the scan should be discussed with the patient and documented. If there are concerns that the scan is not normal, then the patient should be referred directly for specialist fetal echocardiography.

BCCA Fetal Cardiology Standards (September 2021)

These standards are designed for paediatric cardiology tertiary centres and for fetal medicine units offering a fetal cardiology service or echocardiography. These standards are not aimed at obstetric ultrasound departments performing the initial screening of the fetal heart during the anomaly scan. The document has two sections:

Section 1 - Standards for a Tertiary Fetal Cardiac Unit

- A** - Infrastructure of the fetal cardiology unit - staff, equipment, image storage, safety
- B** - Indications for patients to be seen in Fetal cardiology, time needed for scan, recommendations for repeat scan in high-risk patients and perinatal planning
- C** - Information and support for those with an abnormality: clinical nurse specialist/palliative care/psychology/parent support groups. Support for parents deciding on termination of pregnancy
- D** - The Fetal Cardiology Network – Fetal medicine, obstetrics, multidisciplinary meetings, second opinions

Section 2 – Standard for detailed fetal echocardiography in patients at increased risk of congenital heart disease.

Guidance on Fetal echocardiogram in patients that meet the indication for detailed Fetal echocardiography due to maternal or Fetal risk factors or increased risk at first trimester screening

[Microsoft Word - vj_28th Sept_BCCA STANDARDS FETAL CARDIOLOGY_Sept 2021.docx \(bcca-uk.org\)](#)

Referral in North-West, North Wales & the Isle of Man

In the North-West, North Wales and the Isle of Man there are two specialist fetal medicine units providing a fetal cardiology service in conjunction with paediatric cardiologists working across the network. These are:

1. Liverpool Women's Hospital in Liverpool (nearest to surgical centre) and
2. St Mary's Hospital in Manchester

This involves a multidisciplinary approach to include fetal cardiologists, fetal medicine specialists, neonatologists, specialist midwives, specialist fetal cardiac nurse specialists and sonographers. The system for referring patients across the region is different between England and North Wales.

Referral to fetal cardiology in England

Should a sonographer performing a 20-week scan suspect a cardiac anomaly the patient is referred to the local fetal medicine unit for a fetal echocardiogram performed by a fetal consultant cardiologist. It is advised that this referral is made urgently and should not be delayed by local review by a consultant. All urgent referrals to be accompanied by a phone call.

Fetal Medicine Unit in Manchester	Fetal Medicine Unit in Liverpool
<p>St Marys Hospital Tel 0161 276 6385 Email: fetalmedicine.orc@mft.nhs.uk</p>	<p>Liverpool Women's Hospital Tel: 0151 702 4072 Email: lwft.fetalcentre@nhs.net</p>

Referral to fetal cardiology in Wales

There are three hospitals in North Wales that may refer patients to the Liverpool Women's Hospital for detailed fetal cardiology assessment. These are Glan Clwyd Hospital, Rhyl; Ysbyty Gwynedd, Bangor and Wrexham Maelor, Wrexham.

- If unable to obtain adequate views on the 20-week scan at either Glan Clwyd Hospital or Ysbyty Gwynedd, the woman is referred first to Wrexham Maelor Hospital for a scan by their fetal medicine consultant.
- If a cardiac abnormality is suspected, then the patient should be referred DIRECTLY to the Liverpool Women's Hospital to be seen in the fetal cardiac clinic by a consultant cardiologist.

The Antenatal Screening Wales Policy, Standards and Protocols can be found [here](#).



It should be noted that laws surrounding termination of pregnancy in England and Wales change at 22+ weeks and therefore early referral (direct from sonographer) is strongly recommended.

Pregnancies at increased risk for fetal CHD

The majority of cases of fetal CHD occur in low-risk pregnancies. These will only be detected during routine screening, usually at the 20-week ultrasound scan.

Pregnancies that are considered at increased risk for fetal CHD are commonly referred to a local tertiary center for fetal cardiology assessment. The risk of CHD is dependent on the individual group. In some cases, this may be offered at 14+ weeks of pregnancy. Most of the cases screened will be normal though the assessment often provides vital reassurance for the family particularly after a severe CHD diagnosis in a previous pregnancy. The list of maternal, fetal and familial indicators that determine an increased risk of CHD can be found in the BCCA (2021) standards.

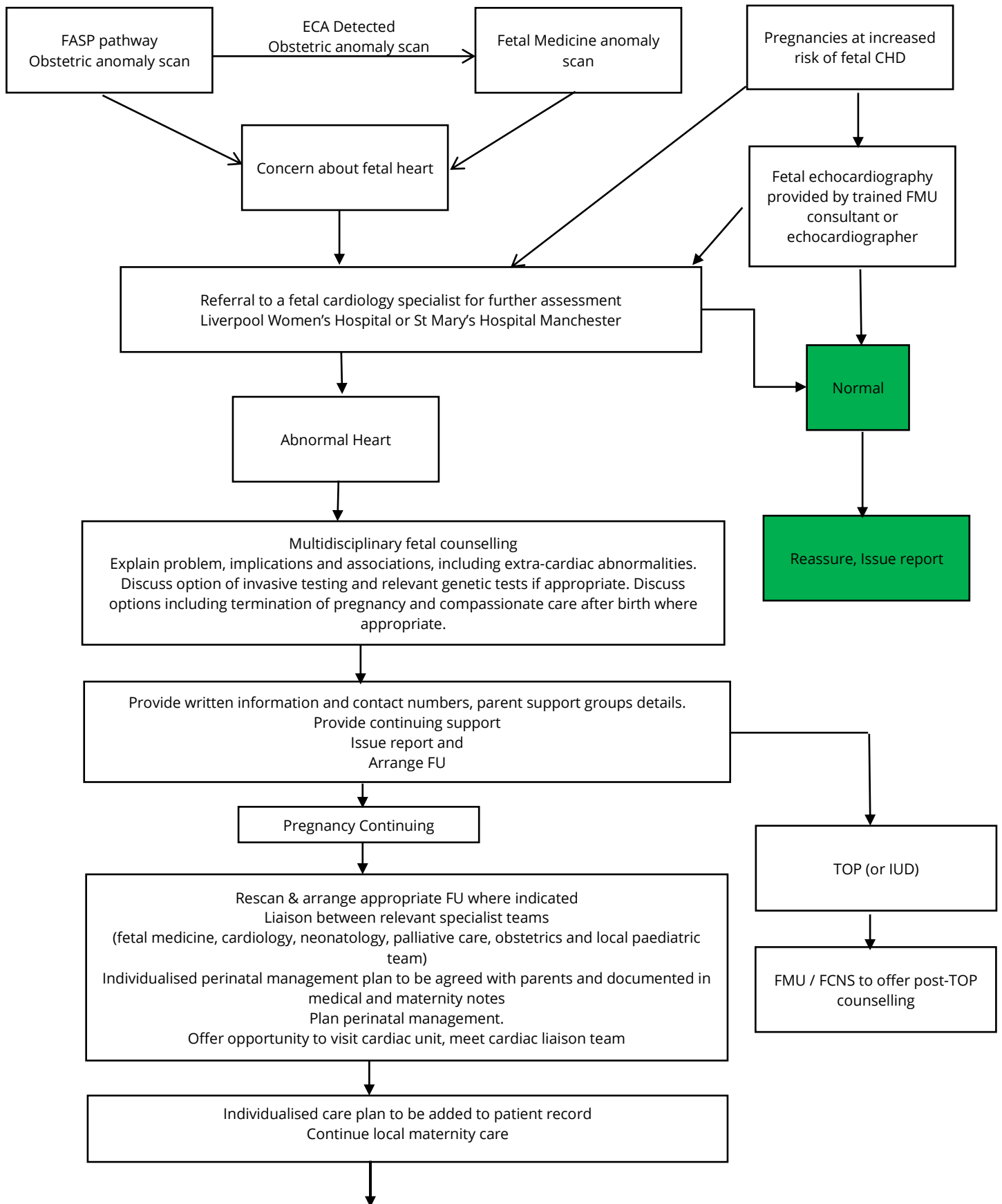
High risk screening scans may be performed by trained professionals such as sonographers or cardiac physiologists working within the fetal cardiology service or by fetal medicine consultants. Scanning the fetal heart is complex and occasional practice should be avoided, complete assessments should be made according to the BCCA 2021 standards in section 2.

Fetal referral pathway where diagnosis of CHD is suspected, or fetus is at increased risk of CHD

See diagram below.



Pathway for Fetal Diagnosis of Congenital Heart Disease



Standards and Quality Indicators

All patients referred with suspected fetal CHD should be *offered* an appointment by fetal cardiology specialist within 3 calendar days of the referral being sent

All patients should be seen by a fetal specialist nurse on the day on diagnosis



Following a diagnosis of CHD (see pathway on page 10)

The vast majority of CHD seen in the fetus represents a spectrum and care must be individualised. Fetal cardiac assessment, counselling and management should occur in line with the BCCA Fetal Cardiac Standards and families will be supported by a multi-disciplinary team.

Following initial diagnosis, usually a detailed scan to check for any extra cardiac anomalies will be offered and perinatal planning should begin early with the family. Final plans should be confirmed typically later in gestation between 28 and 34 weeks and documented clearly in the maternity notes. Local obstetric care is essential and should continue throughout pregnancy. Communication with all professionals involved is vital.

Perinatal planning at delivery location (see diagram 2)

Delivery planning will take into account many factors including the particular cardiac condition and physiology as assessed on fetal echocardiogram, parental preference, family circumstances and local neonatal care provisions. The fetal cardiologist or fetal medicine specialist will lead perinatal planning in collaboration with the obstetrician, neonatologist, and family. An effort is made to deliver care locally where possible.

In some cases, the women may need to be delivered at the tertiary neonatal centre or at Liverpool Women's hospital (due to proximity to Alder Hey Children's Hospital). In severe cases that may require immediate intervention transfer out of region to a co-located centre may be required.

A list of Neonatal Intensive Care Units, Local Neonatal Units and Special Care Units can be found [here](#)

Guideline for the perinatal management of the fetus antenatally diagnosed with cardiac disease (CHD)

Clinicians should read the fetal cardiology reports and follow the Network Cardiac Perinatal Management Plan

Duct dependent lesions

- Reference should be made to whether intravenous prostaglandin infusion should be commenced (all these infants should plan to deliver in a Neonatal Intensive Care Unit).
- Initial dose should be 5 nanograms/kg/minute in antenatally diagnosed patients if no clinical concerns. Higher doses may be needed in those with haemodynamic compromise.
- Contact the relevant cardiology team to inform them of the baby's birth and arrange appropriate review.



- A digital referral should be sent ahead of all referred patients – this should contain referring paediatric consultant, patient name, address, DOB and NHS number. Mother's name and patient's GP details (please send a network urgent referral proforma to the relevant centre).
- Send a copy of the fetal cardiology report along with the baby.
- Ensure the mother gets an obstetric appointment with their local team from 30+ weeks in case of early delivery that occurs locally. Ensure the antenatal management plan is highlighted/escalated/put in the correct place in their own notes in case of unexpected delivery locally.
- If transport is required liaise early with the relevant team.

Patients with transposition of the great arteries (with or without VSD)

- All patients likely to require a balloon atrial septostomy (BAS) i.e. transposition of the great arteries with intact ventricular septum should be delivered at Liverpool Women's Hospital.
- Please refer to the NWNODN guideline for the Management of Transposition of The Great arteries. [TGA without a VSD \(NWNODN Document\)](#)
- Ask the parents to come to Alder Hey where possible or note relevant phone numbers, they may be required to consent for an emergency procedure.

Patients with suspected coarctation of the aorta (CoA)

Coarctation of the aorta remains a challenging diagnosis to make prenatally with a considerable false positive rate and low prenatal detection rates.

- Following fetal cardiology review suspicion of CoA is usually rated as low, medium or high, some infants may be routinely started on prostaglandin.
- For others the local unit will be asked to monitor femoral pulses, pre and post ductal saturations and neonatal condition (intravenous prostaglandin to be started if there are concerns). Regular scans may be required to assess the arch and are required until the duct closes to exclude the diagnosis.
- Patients with a prenatal suspicion of CoA should be followed up until around 12 months (even in the presence of a normal neonatal echo) as later coarctation has been reported.

Non-duct dependent lesions

CHD expected to be hemodynamically stable in the neonatal period

- Those babies with non-duct dependent lesions are expected to be well after delivery and can deliver in their Local Neonatal Unit
- Any neonate who is unwell should be discussed urgently with the relevant cardiology team
- Neonates with an uncomplicated AVSD or Tetralogy of Fallot should be observed on the Neonatal Unit for a minimum of 48 hours following delivery. Oxygen saturations should be monitored continuously for 24hrs followed by regular monitoring of oxygen saturations which could take place on the Neonatal or Transitional Care Unit
- All patients should be discussed with cardiology as soon as possible (preferably within working hours) and an Urgent Advice Proforma completed, and parents should be given an appointment date for cardiology review prior to discharge from hospital. The urgent Advice Proforma can be downloaded from the CHD Network website [here](#)



Prenatal diagnosis that may represent a normal variation

These infants can deliver in their local unit. Care plan and fetal cardiac reports should be available to the team. If the baby is well the normal newborn examination will suffice and they can be discharged with an outpatient referral made to the paediatric cardiology or Paediatrician with Expertise in Cardiology (PEC) clinic for review in the first 2 – 3 months after birth.

- **Right or double aortic arch** – In many infants this will be a variation of normal and won't be clinically significant postnatally. In around 8% there may be associated chromosomal or genetic abnormality, other findings postnatally should be investigated with a microarray in the first instance. A proportion may develop symptoms of a vascular ring with either noisy breathing or difficulty swallowing which should prompt urgent review and investigation.
- **Left Superior Vena Cava (LSVC)** this is often a normal variant and does not cause any cardiac issues as de-oxygenated blood returns to the right atrium via the coronary sinus.

Postnatal genetic testing

- As part of primary postnatal assessment, it is important to make a holistic assessment of the infant and send appropriate genetic tests.
- Please note that the family may have opted to have prenatal genetic testing and these results may be satisfactory to exclude relevant genetic conditions.
- In infants with no clinical signs of genetic syndromes and a negative pre-natal microarray, post-natal genetic testing may not be necessary.
- At present if there is any doubt regarding the appropriate test or if review is required, please contact the clinical genetics team.

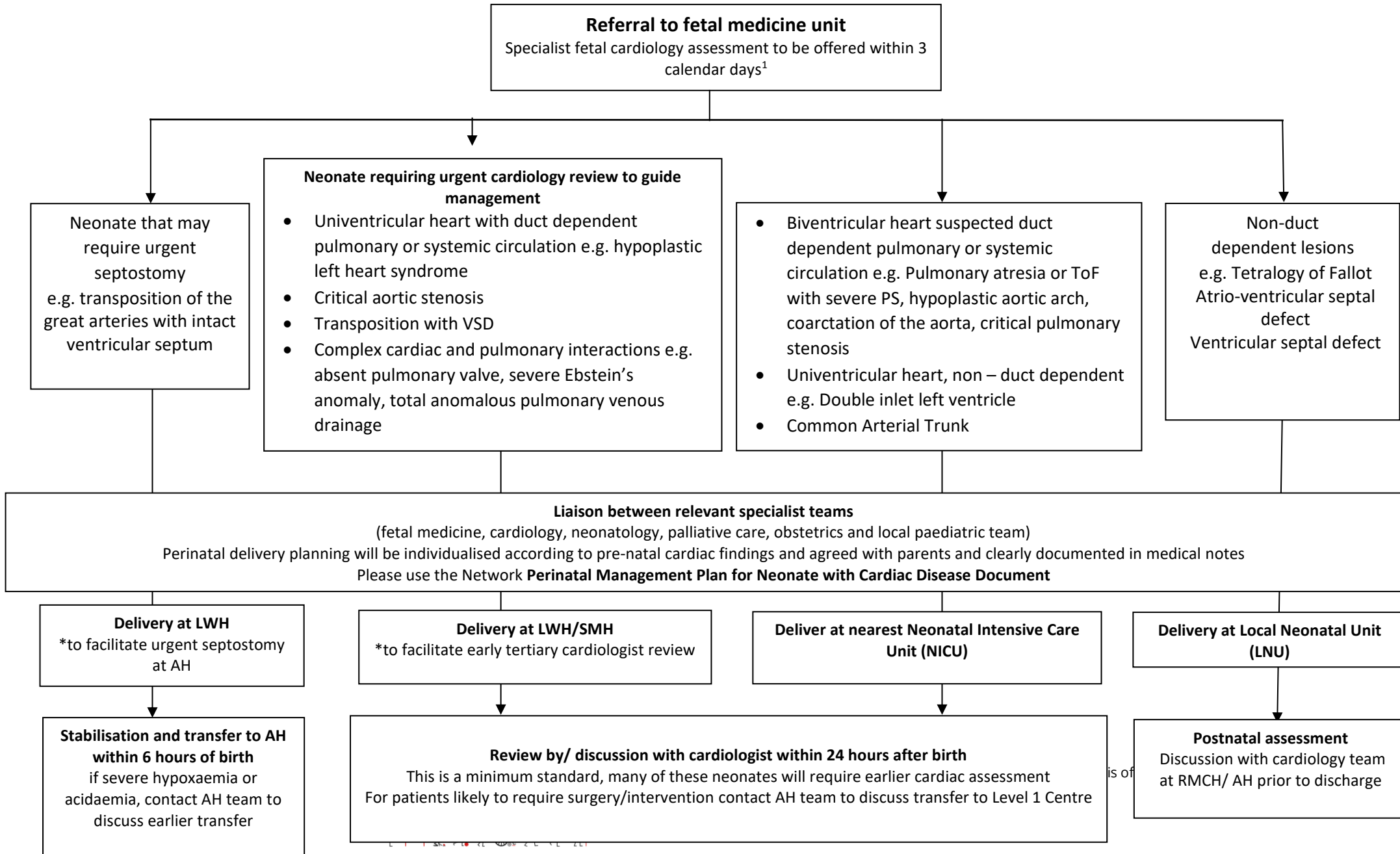
Referral Guidelines

Communication of the management plan

- **All patients with a fetal diagnosis of CHD should have a 'Perinatal Management Plan for Neonate with Cardiac Disease' proforma completed.**
- This is intended to act as an alert for the neonatal and/or paediatric team and outline the basic initial management and signposting to important findings. ***It does not substitute*** for reading the fetal cardiology/ medicine reports where detailed information is recorded.
- The form should be completed by the fetal medicine team and sent to the screening midwife and neonatal +/- paediatric contact at:
 - > The hospital where delivery is planned.
 - > The hospital where the patient was originally booked (in the event of unexpected delivery)
- All teams in the NW CHD ODN are encouraged where possible to have a confidential team mailbox (rather than individual emails) where assurance can be provided, that information can be received and consistently processed.
- At the receiving hospitals the proforma and reports should be:
 - > Entered into the digital or paper maternal notes – labelled where possible as the antenatal management plan.
 - > Sent to the neonatal team as an alert to the upcoming delivery and plan
 - > Where possible digital alerts in the maternal electronic patient record should be created



Diagram 2: Framework for perinatal management of the fetus with CHD



Important Contacts Details

The Fetal Cardiology Teams:

Liverpool Women's Hospital Switchboard 0151 708 9988

- > Fetal Medicine Unit: 0151 702 4072
- > Fetal Cardiac Nurse Specialists 0151 252 5642
 - > Gill McBurney, Marie Murphy
- > Dr Joyce Lim *Consultant Fetal Cardiologist*
- > Dr Ram Ramaraj *Consultant Fetal Cardiologist*
- > Dr Devender Roberts *Clinical Lead Fetal Medicine*
- > Dr Umber Agarwal *Consultant in Fetal Medicine*

St Mary's Hospital, Manchester Switchboard 0161 276 1234

- > Fetal Medicine Unit: 0161 276 6385
- > Paediatric Cardiac Nurse Specialists 0161 701 0664
- > Dr Caroline Jones *Consultant Fetal Cardiologist*
- > Dr Nadezhda Dzhelepova *Consultant Fetal Cardiologist*
- > Dr Louise Simcox *Clinical Lead Fetal Medicine*
- > Dr Sarah Hamilton *Clinical Lead Obstetrics*
- > Lyn Kirby and Della Carr *Lead FMU midwives*

Alder Hey Children's Hospital Switchboard 0151 228 4811

- > On call registrar bleep 369
- > Consultant of the week 0151 252 5220
- > Cardiac secretaries 0151 252 5711
- > Ward 1C (cardiac unit) 0151 252 5418
- > PICU 0151 252 5242
- > Paediatric Cardiac Nurse Specialists 0151 252 5291

Royal Manchester Children's Hospital Switchboard 0161 276 1234

- > RMCH Cons Phone 0161 701 7474
- > Cardiac secretaries 0161 701 2179
- > Ward 85 (cardiac unit) 0161 701 8500
- > PICU 0161 701 8000
- > Paediatric Cardiac Nurse Specialists 0161 701 0664

Transport Teams

- > **Connect North West** (Neonatal transport team) 0300 330 9299
- > **NWTS** (Paediatric transport team) 08000 848 382



References

1. [Fetal anomaly screening programme handbook - GOV.UK \(www.gov.uk\)](http://www.gov.uk)
2. **British Congenital Cardiac Association Fetal Cardiology Standards** (developed by the BCCA Fetal Cardiology Standards working group September 2021): [Microsoft Word - vj_28th Sept_BCCA STANDARDS FETAL CARDIOLOGY Sept 2021.docx \(bcca-uk.org\)](http://www.bcca-uk.org)
3. **Congenital Heart Disease Standards and Specifications. NHS England 2016** [Congenital-heart-disease-standards-and-specifications.pdf \(england.nhs.uk\)](http://www.england.nhs.uk)
4. **Antenatal Screening Wales Policy, Standards and protocols 2023** [Antenatal Screening Wales - Public Health Wales \(nhs.wales\)](http://www.nhs.wales)

