



BRITISH CONGENITAL CARDIAC ASSOCIATION

# British Congenital Cardiac Association Fetal Cardiology Standards

Developed by the British Congenital Cardiac Association (BCCA) Fetal  
Cardiology Standards Working Group  
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## INTRODUCTION

Since the 4-chamber view was introduced into the UK screening programme in the mid 1990s, fetal cardiology has developed and evolved as a specialty to become an integral part of congenital heart disease (CHD) pathways at tertiary congenital cardiac centres. Fetal cardiology is provided by paediatric cardiologists with core training in paediatrics and neonatology, specialty training in paediatric cardiology and sub specialty training in fetal cardiology. This is incorporated into the paediatric cardiology training curriculum as a recognised subspecialist area (1).

As a result of expansion of views used in the UK fetal anomaly screening programme and focussed training of sonographers, antenatal detection rates continue to rise. Fetal cardiac involvement in a wider variety of fetal conditions is also increasingly recognised and requires expert input. Fetal cardiology services have increasingly become the gateway to lifelong care for children with congenital heart disease and our first encounter with these families. It is therefore essential that these services are adequately staffed and resourced to provide state of the art diagnostic facilities and the highest standard of care and support for our patients.

This document contains two sections. The aim of section 1 of this document is to provide an updated framework for fetal cardiologists working in a tertiary fetal cardiology unit following the previous BCCA standards published in 2010 and 2012 (2). The standards within this document are advisory and sit along side the requirements specified for fetal cardiology in the NHSE congenital heart disease standards and specifications (3).

This updated document also includes an additional section, section 2, to approach the question of 'what constitutes a fetal echocardiogram'. This section provides guidance on the level of detail that would be expected in a detailed cardiac scan for a patient with one or more risk factors placing them at higher risk of having a baby with a congenital heart defect.

The service provided by the fetal cardiology unit is to a network of obstetric units linked by the tertiary (level 1) cardiac centre. It is recognised that around the UK some fetal cardiology services are located in a paediatric cardiology unit whilst others are collocated with fetal medicine. Additionally, depending on local resources, a fetal echocardiogram in patients at increased risk of CHD is not always undertaken by a fetal cardiologist. Whilst this section describes the level of detail expected in a fetal cardiology setting for this group of patients it may also act as a guide to those performing a fetal echocardiogram outside the setting of a fetal cardiology service although is not intended as a replacement for other speciality specific guidance.

## OVERVIEW

### **Section 1 - Standards for a Tertiary Fetal Cardiac Unit**

A- Infrastructure of the fetal cardiology unit - staff, equipment, images storage, safety.

- B- Indications for patients to be seen in fetal cardiology, time needed for scan, recommendations for repeat scan in high risk patients, 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.

### **AIMS AND PURPOSE OF A FETAL CARDIOLOGY SERVICE**

- To provide an accurate and detailed diagnosis of congenital heart defects, and acquired or functional heart abnormalities in the fetus.
- To make an accurate diagnosis and initiate treatment, when required, of fetal arrhythmias.
- To provide fast track appointments for suspected abnormalities in line with national standards.
- To provide parents with accurate and evidence-based information on the cardiac abnormality including treatment options, prognosis and further investigations.
- To recognise and arrange further assessment when abnormalities on fetal echo suggest the presence of an extracardiac abnormality.
- To communicate findings to the referring obstetric team and GP and facilitate on going care and referrals to other members of the multidisciplinary team.
- To provide or arrange support for parents following the diagnosis.
- To liaise with referring units to provide optimal perinatal management plans for pregnancies where the fetus has CHD.
- To facilitate transition to postnatal care for ongoing pregnancies.
- To arrange ongoing management and support for patients deciding not to continue with their pregnancy
- To capture, store and archive images on an electronic reporting system.
- To maintain a database to allow audit of outcomes ensuring a high-quality clinical service.

## SECTION 1: STANDARDS FOR A TERTIARY FETAL CARDIOLOGY UNIT

### A: INFRASTRUCTURE OF A TERTIARY FETAL CARDIOLOGY UNIT

#### STAFFING

Each fetal cardiology unit, whether within a tertiary cardiac unit or within fetal medicine, should have a multidisciplinary team comprising doctors, nurses, midwives, and cardiac physiologists specialised in diagnosis and management of fetuses with CHD (2). The team should include:

- 1) **Medical Staff:** The Fetal Cardiologist provides guidance to other clinical staff who perform fetal echocardiograms in their unit including sonographers/physiologists with specialist expertise in fetal cardiology, specialist clinical nurse practitioners, specialist midwives and doctors in training. They will be responsible for the review of fetal echocardiograms when required and provide appropriate counselling.
- 2) **Clinical Nurse Specialists (CNS) in Fetal Cardiology:** The role of CNS in Fetal Cardiology is paramount for the support of the families whose fetus has been diagnosed with CHD. In line with national standards (3), patients should see the CNS within 24 hours of a new diagnosis of CHD. It is preferable that this is face to face at the time of the clinic, but a telephone/video consultation should be offered if that is not feasible. Contact details should be given at each appointment.
- 3) **Specialist Fetal Cardiac Sonographers:** some units may be supported by cardiac physiologists with specialist training in fetal echocardiography.
- 4) **Administrative Staff:** staff are needed to support the fetal cardiology services by receiving referrals, booking appointments in timely manner and ensuring that reports and letters are sent to the relevant teams. Good communication between the relevant teams is important and fetal cardiology reports should be sent to the expectant mother, local obstetrician, GP and midwife.
- 5) **Allied specialties:** there should be access to fetal medicine, midwifery services, genetic services, neonatology, and psychology

#### BASIC REQUIREMENTS FOR FACILITIES, EQUIPMENT AND SAFETY

##### Equipment:

Imaging of the fetal heart and diagnosis of CHD can be challenging because of multiple factors (for example, small cardiac structures, sub-optimal fetal position, maternal habitus and tissue

characteristics). In order to optimise the quality of imaging, high standard ultrasound equipment is required for fetal echocardiography.

The ultrasound system, as a minimum, should have 2-Dimensional (2-D) or grey scale, colour, pulsed-wave Doppler, continuous wave Doppler and M-mode facilities (4). High and low frequency transducers should be available to allow for imaging from first trimester to late gestational age and to adjust for patient's characteristics. The highest possible transducer frequency that allows the combination of best resolution and adequate penetration should be used for each patient (5).

The settings of the system should be optimised so that the highest possible frame rate is provided (4, 5). The images should be magnified such that the relevant cardiac structures can be adequately visualised and cine-loop features are important for real-time assessment of cardiac structures (5).

All professionals scanning should be aware of good ergonomic practice and avoidance of musculo-skeletal injury should be prioritised. Chair, patient bed and keyboard height should be adjustable to maintain optimum ergonomic posture. Chair should have appropriate configuration for ultrasound scanning.

Appropriate training and access to occupational health expertise should be available to facilitate this. The size of the room should accommodate a good ergonomic setup.

**Image storage:**

According to the NHS Fetal Anomaly Screening Programme, still ultrasound images should be captured, stored and archived on an electronic reporting system and there should be a permanent electronic record of all imaging studies which should be accompanied by an electronic report available with the images (6).

In fetal echocardiography digital cine clips are essential for diagnosis of congenital heart disease and therefore units undertaking these scans should have facilities in place to record and store a digital moving image for a complete maternal record.

**Clinic Template:**

Should allow time to complete a specialist fetal cardiac scan and provide counselling to avoid patient delay.

**Safety:**

All health professionals performing fetal echocardiograms should be aware of the Royal College of Radiologists (RCR) and Society and College of Radiographer's (SCoR) standards (6) and they should adhere to the British Medical Ultrasound Society (BMUS) recommended scanning time limits for obstetric scanning (6, 7). The ALARA principle (as low as reasonably achievable) should be applied especially for fetal echocardiograms performed at earlier stages of gestation.

## B: INDICATIONS FOR FETAL ECHOCARDIOGRAPHY

The population-based incidence of CHD in live newborns is 0.6-1.2%. There are a variety of known associations of CHD such as sonographic findings, pre-existing conditions in the expectant mother or congenital heart disease in the family that may increase this risk.

The presence of any factors which represent an increase in this risk should lead to consideration for fetal echocardiography. The level of risk and availability of local provision should be used to determine whether a patient is offered fetal echocardiography locally or referral to a tertiary fetal cardiology service.

**The highest risk group are those where congenital heart disease is suspected at an ultrasound examination and these cases should be referred to the fetal cardiac unit for urgent evaluation.**

The detailed fetal echocardiogram should be undertaken according to the standards described in section 2.

### FETAL INDICATIONS

#### 1. Suspected congenital heart disease

#### 2. Suspected fetal arrhythmia

- FHR >180 bpm
- FHR <120 bpm
- An irregular fetal heart rate can be managed in conjunction with the local obstetric team if a local protocol exists for atrial ectopic beats

#### 3. Increased nuchal translucency

The risk of congenital heart disease increases with increasing NT >95<sup>th</sup> centile (8). The NT threshold for specialist fetal echocardiogram varies amongst fetal medicine and fetal cardiac units. Commonly used thresholds include: NT >95<sup>th</sup> centile, NT >99<sup>th</sup> centile, NT > 3.5mm and the timing of the fetal echocardiogram may vary according to the unit's capacity and expertise. For example, some units advocate for a fetal echocardiogram <16weeks when NT >99<sup>th</sup> centile and when NT 95-99<sup>th</sup> centile, the fetal echocardiogram is undertaken at 19-23 weeks gestation.

We recommend that NT greater than or equal to 3.5mm which approximates to >99<sup>th</sup> centile as an indication for fetal echocardiogram. To facilitate patients obtaining as much information as possible at as early gestation as possible the fetal echocardiogram would ideally be offered by 16 weeks gestation.

Risk factor: fetus	Risk of Congenital heart disease (%)
<b>Nuchal translucency (Hyett et al (8))</b>	
<95 <sup>th</sup> centile	0.08
≥ 95 <sup>th</sup> centile to 3.4mm	0.53
3.5 to 4.4mm	2.89
4.5 to 5.4mm	9.09
≥ 5.5mm	19.51
≥8.5mm	>60%
<b>Extracardiac anomalies</b>	
Hydrops	15-25%
Pleural/pericardial effusions	Variable
Ascites	Variable
Polyhydramnios	Variable
Gastrointestinal malformations eg atresia, congenital diaphragmatic hernia, exomphalos	Varies with anomaly <30%
Neurological malformations	Variable
Lung malformations eg CPAM	Low
<b>Twin pregnancies</b>	
Monochorionic twins	2-10%
Dichorionic diamniotic (DCDA) twins	Low
<b>Fetal 1<sup>st</sup> trimester sonographic markers</b>	
Tricuspid regurgitation in a low risk population (NT<95 <sup>th</sup> centile)(9)	Low
Reversed flow in the ductus venosus	Low in isolation, 15% if NT>95 <sup>th</sup> centile in euploid fetuses
Aberrant right subclavian artery	Low
<b>Fetal conditions associated with fetal heart failure</b>	
Anaemia	
Tumours with large vascular supply	
Arteriovenous malformation	
Absence of the ductus venosus	Also, Noonan syndrome-associated CHD
Acardiac twin pregnancy	
Twin to twin transfusion syndrome	~10%

## MATERNAL INDICATIONS

### Family history of congenital heart disease

- This is usually restricted to cases where a first degree relative of the fetus has a diagnosis of CHD (in the absence of underlying chromosomal or genetic abnormality):
  - Mother: 3-7%
  - Father: 2-3%
  - Affected sibling: 3%

- Risk may be higher for left-sided obstructive lesions (10, 11) up to 8% for recurrence of HLHS when a previous child affected have been reported. It should be noted that this may be overestimated as a higher risk of recurrence is reported from studies that have include family members with lesions such as bicuspid aortic valve.
- Risk increased if > 1 family member is affected  
Unless multiple family members affected, referral is not indicated for 2<sup>nd</sup> degree relative with CHD (risk 1-2% to fetus) or 3<sup>rd</sup> degree relative with CHD (~ 1% risk to fetus)
- This excludes those conditions that are present as obligatory shunts in the fetal circulation such as atrial septal defect (ASD) and patent arterial duct (PDA) and first degree relative with mitral valve prolapse.
- First degree relative with cardiomyopathy, particularly those presenting in utero or in infancy.

Risk factor: maternal	Risk of CHD (if known)
Maternal congenital heart disease*	2-7%
*This excludes those conditions that are present as obligatory shunts in the fetal circulation such as atrial septal defect and patent arterial duct. *risk may be >6% in left-sided obstructive lesions	
Maternal diabetes	
Pre-conception diabetes	3-5%
Poorly controlled maternal DM in 1 <sup>st</sup> trimester	3-5%
Gestational diabetes	<1%
Insulin resistance 3rd trimester	<1%
Maternal anti-Ro/SSA or anti-LA/SSB antibodies	
<i>These can be present in systemic lupus erythematosus, Sjogren's syndrome</i> <i>Maternal antibody titres may reflect fetal risk</i>	
Background	1-5%
Previous affected child	11-19%
Maternal phenylketonuria	
Risk of CHD when poor maternal control periconception of phenylalanine concentration >10mg/dL	12-14%
Maternal medications	
Sodium valproate, carbamazepine	1-2%
Other anti-convulsants	
Lithium	1-2%
Retinoic acid	> 2%
Non-steroidal anti-inflammatory drugs in 1 <sup>st</sup> /2 <sup>nd</sup> trimester	1- 2%
Non-steroidal anti-inflammatory drugs in 3 <sup>rd</sup> trimester	Reversible constriction of arterial duct
Chemotherapeutic agents	Variable with agent
Methotrexate	



Selective serotonin reuptake inhibitors (SSRI) – paroxetine only	1-2%
Other selective serotonin reuptake inhibitors (not paroxetine)	<1%
ACE inhibitors eg lisinopril	> 2%
Maternal viral infection	
Rubella in 1 <sup>st</sup> trimester	>2%
Maternal viruses associated with fetal myocarditis eg coxsackie, CMV, toxoplasma, parvovirus	
IVF Conception (12)	1-3.3%

### Inadequate Views

Where the cardiac components of the anomaly scan could not be completed, FASP guidance recommends that the screening anomaly scan should be repeated within 2 weeks, if the cardiac views cannot be completed then the limitation of the scan should be discussed with the patient and documented.

If there are concerns of CHD, rather than difficulties visualising, then the patient should be referred for specialist fetal echocardiography.

### TIME REQUIRED FOR SCAN

The suggested time needed for a detailed assessment of the fetal heart is 30-45 minutes, this includes the time for explanation of the limitations of fetal heart scanning, preparation of the patient and conveying a normal result. When congenital heart disease is identified, additional time is required for counselling. Multiple pregnancies may also require additional scan time.

### TIMING OF SCAN

	Timing of appointment
Suspected fetal cardiac abnormality	<ul style="list-style-type: none"> <li>In line with NHS national guidance, currently within 3 calendar days of referral (3)</li> <li>It would be considered good practice to assess a fetal tachyarrhythmia within 24 hours. A local pathway for out of hours/ weekend presentation should be available.</li> </ul>
Increased risk of CHD	<ul style="list-style-type: none"> <li>18-22 weeks ideally to allow time to obtain result from invasive test if abnormality detected.</li> <li>Family history of major CHD may be seen in first trimester (12-14 weeks) where this can have an additional important role in parental reassurance</li> <li>NT &gt;99<sup>th</sup> centile (ideally 12-16 weeks)</li> </ul>

## RECOMMENDATION FOR REPEAT ECHOCARDIOGRAM

In some cases, congenital heart disease may evolve or increase in severity as pregnancy progresses and warrant a further examination at later gestation or postnatally. This includes coarctation of the aorta, aortic stenosis, pulmonary stenosis, cardiomyopathy and complete heart block. In pregnancies, where there is a first-degree family relative with these conditions a repeat echocardiogram may be offered. If local resources do not allow for a repeat scan, it is important to inform the expectant women, the limitations of the mid-gestation fetal echocardiogram.

### Indications for repeat echocardiogram following a normal echocardiogram at 20 weeks

- Family history of left sided obstructive lesions including classical HLHS, coarctation of the aorta and aortic stenosis.
- Maternal anti-Ro/La antibodies
- Persistent elevation of nuchal fold thickness (>6mm)
- First degree relative (eg previous sibling) with cardiomyopathy presenting in infancy

Expectant women who are known to have anti-Ro antibodies and have had a normal 18-20 week fetal echocardiogram a repeat assessment of fetal heart rhythm is recommended between 26-28 weeks' gestation. Surveillance between appointments should be provided by auscultation of the fetal heart at midwifery appointments or in combination with fetal cardiology if resources available.

A small proportion of fetuses with NT > 95<sup>th</sup> centile have a persistent increase in the nuchal thickness at 20 weeks (nuchal fold thickness). This can be associated with Noonan syndrome and evolution of cardiac findings such as valvar stenosis and hypertrophic cardiomyopathy. A third trimester fetal echocardiogram is recommended in such circumstances.

### Early Fetal Echo

All patients seen at < 16 weeks should have a repeat assessment at 20 – 22 weeks

Patients seen at 16-18 weeks normally warrant second scan later in pregnancy unless excellent imaging obtained at primary assessment

## PERINATAL PLANNING FOLLOWING THE DIAGNOSIS OF FETAL CARDIAC ABNORMALITY

### Timing of delivery

The aim should be, in the absence of other fetal or maternal concerns requiring early delivery, to deliver the fetus with CHD as close to term as possible, to avoid any additional risk of prematurity. Previous studies have demonstrated an adverse impact of early delivery on surgical and neurodevelopmental outcome (13, 14).

It may sometimes be necessary to recommend induction of labour, or planned C section, to facilitate early perinatal management and reduce the risk of delivery outside of a specialist hospital setting.

The risk and benefit of management options should be discussed with the parents by the obstetric and fetal cardiology team.

### **Location of delivery**

This will be determined by local network arrangements and in discussion with the booking maternity units. The appropriate unit will depend on the risk of early decompensation with the heart condition and the local provision and confidence in the management of neonates with congenital heart disease

We would recommend that particular consideration is given to delivering the following conditions at a unit in close proximity to the tertiary cardiac centre.

<b>Condition</b>	<b>Indication for delivery at or close to cardiac centre</b>
Transposition of the great arteries with intact ventricular septum	Fetal cardiology can stratify the risk in fetuses with TGA (15). Despite this, a small proportion of patients with TGA present with severe cyanosis in the immediate newborn period requiring emergency balloon atrial septostomy.
Hypoplastic left heart syndrome with evidence of restriction of the interatrial septum	In the presence of a restrictive atrial communication there may be an acute increase in left atrial pressure when pulmonary venous return increases with the fall in pulmonary vascular resistance after birth. This can lead to acute left atrial hypertension, pulmonary oedema and low cardiac output resulting in death.
Complete heart block	Urgent chronotropic support or pacing may be required after birth to achieve haemodynamic stability
Uncontrolled tachyarrhythmia	Haemodynamic compromise may result from sustained tachycardia. Delivery near a cardiac centre will facilitate early assessment of the newborn with intensive care support and options of cardioversion or pharmacological therapy after birth if needed.
Total anomalous pulmonary venous connection (TAPVC)	If there is concern that the pulmonary venous drainage may become obstructed in the early newborn period for example any case of infracardiac TAPVC or evidence of restrictive atrial communication delivery

	close to the cardiac centre would be recommended.
Other high-risk conditions may include tetralogy of Fallot with absent pulmonary valve, cardiac tumours, Ebstein's anomaly of the tricuspid valve with severe tricuspid regurgitation/cardiomegaly	

Where high risk patients do not live in close proximity to the tertiary cardiac unit consideration should be given to providing an alert card to expedite assessment in maternity/labour ward triage should they present at their local unit in early labour or with pregnancy complications.

Separation of mother and baby – where delivery cannot take place at the cardiac centre, consideration should be given to minimise the impact of separation of mother and baby in the newborn period.

### **Neonatal Transfer**

If patients are delivered at a site not collocated with the cardiac unit there should be a clear perinatal plan in place for transfer, by a team with specialist experienced in the management of babies with congenital heart disease, with support of the local cardiac intensive care/ cardiology team.

## **C: INFORMATION AND SUPPORT FOR PARENTS**

### **Before the scan**

1. Information leaflet should be sent with appointment details (or departmental website information) outlining location of clinic, what to expect during the scan and who can attend appointment.
2. Additional support should be considered and provided if required for those with limited English language and those with physical, sensory or learning needs for example: foreign language interpreting service, face to face wherever possible; British Sign Language (BSL) interpreter or support worker.
3. Information should be given regarding consent for scan and data collection

### **At the time of the scan**

1. Ensure parents understand the reason for specialist fetal cardiac evaluation, for example an abnormality has been suspected or deemed high risk.
2. Explain what to expect during scan; it may take some time to gather all the information, and then the scan results will be discussed in a separate room.

3. Explain the limitations of a fetal echocardiogram due to the unique fetal circulation.

### **Counselling following the identification of an abnormality**

Following the detection of a problem, information, counselling and support should be provided.

1. Provision of information with implications and choices for fetal heart abnormalities should be provided by a fetal cardiologist or paediatric cardiologist with experience of fetal congenital heart disease.
2. A separate room should be available for discussion of the scan findings. This should be in close proximity to the scanning room to provide a quiet environment, minimising the potential for interruption (16)
3. Information should be jargon free and delivered in an empathetic and compassionate way (17)
4. Information should include an accurate description of the abnormality, information regarding the need for non-surgical or surgical intervention; potential surgical options available for the condition; timing and number of planned interventions likely to be required; associated mortality and morbidity and the short and longer term prognosis for the child.
5. Discussion and presentation of choices should aim to be in as non-directive manner as possible.
6. When present, the impact of extracardiac structural abnormalities in combination with the cardiac abnormality should be discussed and the potential implications for postnatal management. This may involve joint discussion with the FMU specialist.
7. The potential associations with chromosomal or genetic abnormalities should be discussed and the option of invasive or where appropriate, non-invasive testing, should be offered in a timely manner.
8. The parents should be made aware of all options available to them including, where relevant, information on termination of the pregnancy and relevant time limitations for decision making. Sufficient information and support must be provided to enable them to make an informed decision for their individual circumstances. Pregnancy options should be presented in an unbiased, non-judgemental manner (18)
9. In the presence of certain complex and life limiting congenital heart disease, the option of postnatal comfort care or palliative care should be discussed.
10. A fetal cardiac nurse specialist or specialist fetal medicine midwife should be present during both the primary consultation in accordance with UK National standards. It is also of benefit for the specialist nurse/midwife to be present at follow up consultations. Documentation of information given by the nurse specialist/midwife should be recorded.
11. Clear documentation should be made in the patients records of the scan findings, discussion during the counselling session, appropriate follow up or onwards referral.
12. The individual values and beliefs of patients and the impact of these on decision making should be considered and respected.

### **Written information/resources**

- The cardiologist should provide the parents with a written report summarising the information given during the consultation.
- This can be supplemented with written information on the specific diagnosis including diagrams of the cardiac condition. Signposting to trustworthy, accurate websites may be useful, for example, British Heart Foundation, Childrens Heart Federation or other appropriate resources.
- Contact details should be provided for independent counselling groups, and appropriate patient related support groups. This may include signposting to support groups of relevant faiths when appropriate.
- Parents should be provided with contact names of staff they have met during the consultation and contact details for the fetal cardiac nurse specialist/ specialist fetal medicine midwife who will provide continuing support.
- Written communication regarding the scan findings and consultation discussion should be provided, ideally on the same day, to referring team; GP; patient and other members of the multi-disciplinary team when appropriate

### **Parent support**

- Parents should be given adequate time to process the information that they have been given during the consultation, they should be allowed time to express their grief and be left alone if desired.
- The parents will experience a range of emotions after being told that their baby has a serious heart problem, and this may make it difficult for them understand all the information in one consultation. Support with further information and emotional support is likely to be required. This may be provided by a nurse practitioner/counsellor/specialist practitioner who has been present during the counselling session (19).
- Referral to psychology service can be offered if considered appropriate and this resource is available.

### **Communication with other teams and ongoing care**

1. The family should have appropriate follow up arrangements made.
2. If not already undertaken a referral should be made to an associated feto-maternal medicine unit to identify any extra-cardiac malformations; when appropriate to discuss testing for fetal chromosomal and genetic abnormalities with chorionic villus sampling (CVS) or amniocentesis and more detailed discussion of the option of termination of pregnancy if required.
3. If appropriate, onward referral to psychological services, clinical genetics, palliative care services (20) and bereavement services.
4. Counselling at follow up scans may be undertaken in the presence of feto-maternal medicine obstetrician and/or with other members of the multi-disciplinary team: clinical geneticists; neonatologists; palliative care team or a paediatric cardiac surgeon if appropriate.
5. Cases in which management options are unclear or complex may benefit from multidisciplinary team discussion. In some cases it may be most appropriate that this

is a prenatal MDT involving fetal medicine specialists and fetal cardiologists. In others, particularly where there is uncertainty about the postnatal management course, discussion at a cardiac surgical MDT may be more appropriate. The outcome should be documented and fed back to parents.

6. Discussion should be undertaken regarding the time and place of delivery, for example in local obstetric unit vs. tertiary centre at/close to paediatric cardiology services. This discussion should involve the local obstetric and neonatal teams in addition to the tertiary unit and cardiology team.
7. A meeting, for the parents, with a member of the neonatal team should be considered at third trimester follow up appointments for major CHD
8. Where resources are available, parents may benefit from the provision of antenatal support/ information sessions. These may include information from parents who have previously had a child with a congenital heart defect and practical information such as a tour of the hospital.

### **Bereavement support**

Support and the opportunity for further discussion should be given to parents who have suffered a pregnancy loss whether termination of pregnancy or intra uterine demise. This may be provided by fetal cardiology or, if more appropriate, the fetal medicine team.

## **D: NETWORK, TRAINING, QUALITY ASSURANCE**

### **Network**

The fetal cardiology services should be standardised across the congenital heart disease network. The holistic approach and management of the fetus and expectant mother is paramount for the outcome of pregnancy, post-natal management and longer term prognosis. Good communication between the relevant teams is vital and fetal cardiology reports should be sent to the expectant woman, local team (obstetrician, neonatologist and paediatrician with expertise in cardiology (PEC)), GP and midwife.

### **Training**

A programme of accessible training, which may include educational events, feedback, review and shared learning and training in fetal cardiac imaging, should be provided by the tertiary centre to support the local and aligned network of maternity centres providing routine anomaly ultrasound screening.

There should be an expectation that the service will provide/lead training for paediatric cardiology trainees to achieve curriculum competencies (ST-4 to 6) and special interest (themed for service) training (ST7/8), as well as fetomaternal medicine trainees. Training and updates should be provided for obstetric sonographers and other health care professionals.

### **Quality Assurance**

Tertiary centres should have mechanisms in place for audit and quality improvement. This should include maintaining a database of patients seen in fetal cardiology. Seeking and documenting outcomes to ensure that prenatal diagnosis is accurate.

There should be an internal governance mechanism to discuss fetal cases in which there is diagnostic difficulty. Additionally, there should be a forum to identify and review cases where there are new or unexpected postnatal findings to ensure a high standard of prenatal diagnosis and opportunities for learning from cases where an abnormality is missed

### **Continuing Professional Development**

Clinicians involved in fetal cardiology would be expected to seek feedback of postnatal course including participation in multi-disciplinary morbidity & mortality review within the cardiac centre.



## SECTION 2: GUIDANCE ON PERFORMING FETAL ECHOCARDIOGRAPHY IN PATIENTS AT INCREASED RISK OF CONGENITAL HEART DISEASE

### What Constitutes a Fetal Echocardiogram?

#### Introduction

Patients at increased risk of congenital heart disease (CHD) in the fetus should be offered detailed diagnostic assessment of the fetal heart above that provided in the UK national screening programme (5, 6), this includes 2D imaging, colour flow mapping, pulsed wave Doppler as outlined below.

Fetal echocardiography for pregnancies at increased risk of CHD may be not always be undertaken by a fetal cardiologist, even in centres with fetal cardiac units, for example these patients may be scanned by specialist sonographers or fetal medicine specialists. This is dependent on local resources and expertise. This consensus document aims to give guidance as to the views and the structures that should be visualised to fully assess these high-risk patients. This is not intended as replacement for detailed statements on fetal heart imaging (4), but as a practical reference to ensure adequate detail is obtained. This represents a core minimum dataset. If an abnormality is identified then further imaging and measurements may be necessary for diagnosis and prognostication.

- If as a result of this assessment of the fetal heart an abnormality is identified, the patient should be seen by a fetal cardiologist.
- In the absence of underlying chromosomal or genetic abnormality, termination of pregnancy should not be offered without fetal cardiology assessment to define the diagnosis and provide appropriate counselling.

#### Maintaining skills of personnel

Performing fetal echocardiography requires a specific skill set, knowledge of cardiac anatomy and focused training. On-going learning is needed to develop these skills and to keep up to date. Those providing fetal echocardiography should maintain close links with fetal cardiology colleagues. It is recommended that clinicians or sonographers undertaking this work should be involved in performing these types of assessments on a regular basis and occasional practice is discouraged. It is essential professionals maintain competency by attending educational activities and working alongside fetal cardiology colleagues. Provision should be made in job plans to support this.

#### History

Prior to starting the scan, the following should be established and recorded.

1. Reason for referral
2. Previous pregnancy history
3. History of any relative born with cardiac or congenital defect, including type of CHD
4. First trimester screening result including NT measurement, NIPT result if done.
5. Maternal health and medications

### Documenting findings of fetal echocardiography

- The majority of the fetal cardiac anatomy should be demonstrated by storing cine loops to clearly view cardiac contractility, valve movement and blood flow patterns
- Stored images should all be archived to allow later review and audit
- Reports should detail all structures demonstrated with particular reference to any aspects that were not seen ( guidance list below)

### Making measurements of cardiac structures

- It is good practice to make standardised measurements when assessing the fetal heart
- A number of reference ranges are available, and departments should decide which to use and follow the methodology for measurement eg. systole vs. diastole, inner edge to inner edge vs. leading edge to leading edge.
- Measurements should be z scored according to larger published series given variations in size through gestation (21-23)
- Gestation-specific heart rate and valve Doppler reference ranges / z scores are available (24)
- Ultrasound machines can be programmed by the vendor to automatically provide z scores during live scanning. Software such as View Point will automatically reference measurements against reference data, apps such as CardioZ (cardioz.com) and Parameter z (fetal.parameterz.com) are available to z score fetal data.

There are some conditions, such as anti-Ro/la Ab positive expectant women, who in addition to the standard level 1 screening, may also require specialist assessment of fetal heart rhythm.

## STANDARD VIEWS TO BE RECORDED WITH CORE AND DESIRABLE FEATURES TO IDENTIFY ON EACH VIEW

The scan should begin with careful identification of the fetal position and left/right orientation. Throughout the 5 transverse views the sonographer should remain in, as near as possible, a true transverse cut through the fetal chest.

View	Core features to identify	Desirable features to identify
<u>Situs (transverse view of abdomen)</u>	<b>2D imaging</b> Aorta slightly to left of spine IVC anterior and right of aorta	<i>Colour ductus venosus</i>

	<p>Stomach bubble on left Umbilical vein</p>	<p><i>Pulsed wave Doppler (PWD) ductus venosus</i></p>
<p><b><u>4 chamber view (transverse)</u></b></p> <p><i>*The 4-chamber view should be reviewed in two planes, cardiac apex pointing up towards the probe (good for viewing AV valve differential insertion) and perpendicular to the ultrasound beam (to view the atrial and ventricular septums and wall thickness)</i></p>	<p><b>2D imaging</b></p> <p>Heart size (subjective assessment on view of full fetal thorax)</p> <p>Heart position – look for axis deviation as marker of conotruncal abnormality</p> <p>Balanced ventricular width and length</p> <p>Contractility and wall thickness</p> <p>Morphology of ventricles (moderator band in right ventricle, smooth walled left ventricle)</p> <p>Differential insertion of atrioventricular valves with tricuspid valve lower than mitral valve with crux of the heart clearly demonstrated</p> <p>Foramen ovale flap valve deviated from R to L Intact primum atrial septum and ventricular septum</p> <p>Exclusion of pericardial or pleural effusion</p> <p><b>Colour Doppler</b></p> <p>Equal widths of ventricular inflows, non-turbulent flow, inspect for valve regurgitation</p> <p>Ventricular septal defects (note optimise pulse repetition frequency (PRF) and colour gain for VSD)</p> <p>Pulmonary veins - ensure true connection to left atrium as a misleading impression of normal connection may be seen with veins draining into a confluence (a minimum of one vein from left and right should be documented)</p>	<p><i>PWD pulmonary vein from each side</i></p> <p><i>PWD AV valve inflow</i></p> <p><i>Mechanical AV interval measurement when reviewing patients with anti-Ro / La antibodies (25)</i></p> <p><i>If suspicion of asymmetry then measurement of AV valve annulus-Z score</i></p>
<p><b><u>Left ventricular outflow tract (transverse) 2D imaging</u></b></p>	<p>Examine perimembranous inter-ventricular septum in 2D and with colour – ensure continuity of septum up to aortic valve (viewed with ultrasound beam perpendicular to the intraventricular septum)</p>	<p><i>Measure aortic valve diameter – Z score</i></p>

	<p>Aortic valve leaflets – thin, mobile, flatten against wall in systole</p> <p><b>Colour Doppler</b> – with appropriate PRF for gestation, exclude aliasing across aortic valve</p> <p><b>PW</b> Doppler aortic valve to assess within gestation-specific reference range</p> <p>Fetal heart rate should be documented from one outflow valve</p>	
<p><b><u>Right ventricular outflow tract (transverse)</u></b></p> <p><b>2D imaging</b></p>	<p>Right ventricular outflow tract crossing Left ventricular outflow tract (to exclude transposition of the great arteries)</p> <p>Pulmonary valve leaflets – thin, mobile, flatten against wall in systole</p> <p>Main pulmonary artery branching into left and right confluent branch arteries</p> <p><b>Colour</b> – with appropriate PRF for gestation, exclude aliasing across pulmonary valve</p> <p><b>PW</b> Doppler of pulmonary valve to assess within gestation-specific reference range</p>	<p><i>Measure pulmonary valve diameter - Z score</i></p>
<p><b><u>3 vessel view</u></b></p>	<p>Size of vessels correct (Pulmonary artery&gt;Aorta&gt;Superior vena cava)</p> <p>Single superior vena cava (SVC) noted on right</p>	
<p><b><u>3 vessel and tracheal view</u></b></p>	<p>Aorta and Duct passing to left of trachea (making V shape as they join at the isthmus)</p> <p><b>Colour</b> - non-turbulent flow in same direction in both vessel</p>	<p><i>PW – Duct</i></p> <p><i>Measure distal arch and duct</i></p> <p><i>Often the thymus can be clearly seen in this view</i></p>
<p><b>Sagittal views</b></p>		<p>Superior and inferior vena cava to RA</p> <p>Aortic arch</p> <p>Ductal arch</p> <p>Colour arches</p> <p>Measurement of aortic isthmus</p>

### **Cases when completing a full fetal echocardiogram can be challenging**

It is recognised that, even in the most skilled hands, it sometimes may not be possible to complete a full assessment detailing all the structures outlined above. In some cases, maternal habitus, fetal position, multiple pregnancy will limit views available. Reference should be made to which structures have been seen (and those that have not). Limitations should be explained to the patient. If concerns about abnormality, then assessment by a fetal cardiologist is required

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