



**BRITISH CONGENITAL CARDIAC ASSOCIATION**

## **Fetal Cardiology Standards**

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

**Revised April 2012**

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Original version March 2010

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## **Introduction**

Congenital Heart Disease (CHD) may be identified during fetal life with a very high level of diagnostic accuracy at tertiary centres with an established fetal cardiology programme. For some forms of cardiac abnormality there may be an improvement in postnatal outcome with prenatal diagnosis. Furthermore, early diagnosis during pregnancy will allow parents to consider various options and be prepared for subsequent treatments. In order to deliver a comprehensive high quality service, fetal cardiology units should provide appropriate support and information (before and after the fetal heart examination) and liaise with all relevant specialists and support services, in addition to the essential task of providing an accurate cardiac diagnosis.

Most cases of CHD occur in low risk pregnancies and will only be detected by screening at the time of obstetric ultrasound scans. The concept of prenatal screening for CHD was introduced in the UK over 20 years ago and current national guidelines recommend that the heart (views of the four-chambers and great arteries) should be examined at the time of the obstetric anomaly scan (1,2,3). Despite this, there is a large regional variation in prenatal detection rates of CHD at the time of obstetric screening. Teaching and training in general hospitals has been shown to have a positive impact on the detection of congenital heart disease before birth. The means of achieving a more uniform national standard is under review (3).

The standards outlined in this document are designed for paediatric cardiology tertiary centres offering a fetal cardiology service and are aimed at providing a framework for the development of tertiary services, which can be adapted to fit in with local models of delivery. These standards are not aimed at obstetric ultrasound units performing the initial screening of the fetal heart during the fetal anomaly scan. The NHS Fetal Anomaly Screening Programme (FASP) has set standards for the fetal anomaly scan (3). The BCCA and 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.

Cases where there is evidence of fetal congenital heart disease should be referred to a fetal cardiology specialist for a complete cardiac diagnosis. Assessment of non-cardiac fetal abnormality should be undertaken by a fetal medicine specialist, who may also be able to make initial detailed assessment of the fetal heart in addition to the assessment of other abnormalities. Counselling will need to take into account the extent and implications of all associated abnormalities. The working relationship between fetal cardiology specialists and fetal medicine specialists is extremely important in the management of fetal congenital heart disease

## Standards for fetal cardiology service in a tertiary centre

<b><i>Aims and role of a fetal cardiology service</i></b>	
To accurately establish normality or the presence of CHD in the fetus as early as possible. In the abnormal fetal heart, a tertiary level service is expected to make a full and accurate diagnosis of structural and functional defects and rhythm disturbances. A tertiary fetal cardiology service should also be able to recognise features of the cardiac scan that suggest there may be an extra-cardiac abnormality, even though the heart structure is normal. Close liaison between fetal cardiology specialists and fetal medicine specialists is paramount. An early accurate diagnosis will give parents choice, as well as the opportunity to plan the delivery and postnatal management to try and improve the outcome.	
To provide appropriate counselling and support for parents and families following a prenatal diagnosis of CHD.	
To communicate results to the referring obstetric team, local primary care teams and any other relevant medical personnel.	
To plan as a multidisciplinary team the management of on-going pregnancy in order to try and improve outcome. This should include all personnel likely to be involved in perinatal management.	
To facilitate appropriate referral and discussion for ongoing care for those women and their partners not wishing to continue pregnancy or wishing to continue with pregnancy with the view of compassionate care post delivery.	
To initiate prenatal treatment where appropriate e.g. in fetal arrhythmias and selected structural lesions.	
To maintain a database to enable regular audit of activity and to obtain outcome data and to monitor sensitivity, specificity, false negative and false positive diagnoses.	
<b><i>Basic requirements for a fetal cardiology service</i></b>	
<b>1. Staffing</b>	Dedicated multidisciplinary team trained in the diagnosis and management of fetal CHD
a) Medical staff	Each unit should have designated consultant(s) with a special interest and expertise in fetal cardiology, who have fulfilled the training requirements for fetal cardiology as recommended by the paediatric cardiology SAC (4) or the Association of European Paediatric Cardiologists (5).
	Consultant grade staff, who are appropriately trained in fetal cardiology (4,5), must be available to perform and check scans as necessary and see all cases of abnormality. The consultant must have a clear understanding of the legal framework relating to prenatal diagnosis.
b) Nurse practitioner / counsellor/ specialist practitioner/specialist midwife	A named individual in a supportive role should ideally be present or at least be immediately available to provide help and on-going support to families. There should be consistency in this role so that parents can speak with and meet with the same named individual as and when required throughout pregnancy and at follow-up appointments in the tertiary centre. This named individual should make contact with the pregnant lady's local hospital and community teams to inform personal involved in her care about the fetal cardiac findings and implications.

c) Other clinical staff	The following can perform scans under supervision of a consultant trained in fetal cardiology: i. Sonographers ii. Specialist radiographers iii. Specialist nurse practitioners iv. Specialist midwife practitioners v. Doctors in training
<b>2. Time for scans</b>	A minimum of 45 minutes should be allocated for the consultation and fetal echocardiogram. In cases of abnormality the time required is very likely to be longer, particularly for counselling after the diagnosis, and this must be taken into account when booking appointments. Longer appointment times may also be required for multiple pregnancies.
<b>3. Related services</b>	There must be well established links with the following services:
a) Essential	i. Paediatric cardiology /paediatric cardiothoracic surgical unit ii. Neonatal unit iii. Other paediatric medical and surgical services iv. Feto-maternal medicine unit v. Maternity service
b) Desirable	The following links are desirable: i. Genetics department ii. Adult cardiology service iii. Pathology service
<b>4. Equipment</b>	High resolution equipment will allow earlier and more accurate diagnosis and also quicker evaluation. i. Ultrasound equipment must be of high standard ii. Must be maintained or replaced as necessary
<b>5. Database and image storage</b>	A record must be kept of all scans performed (6). i. Must have database for data entry of all scans ii. Must have system of obtaining and recording outcomes for audit iii. Aim for national database for all fetal cardiac diagnosis. This should link with UK CCAD database for cases undergoing cardiac surgery. iv. Must keep videotape/digital recording of all scans
<b>6. Additional network links</b>	Fetal cardiology centres must have network links with obstetric ultrasound units for audit and development. The tertiary centre should help in providing teaching, training and support for obstetric ultrasound units in their network.
<b><i>Safety aspects of fetal ultrasound</i></b>	
	The safe use of ultrasound is of paramount importance and practitioners should be aware of, and adhere to, agreed national safety standards (7).
	All health professionals involved in fetal heart scanning should adhere to NHS FASP safety standards and guidelines for obstetric anomaly scanning.

<b><i>At what stage should fetal heart scans be performed?</i></b>	
<b>1. In case of a suspected cardiac problem</b>	All cases of suspected fetal cardiac abnormality must be seen by a fetal cardiology specialist within 5 working days of referral and preferably within 2 working days if possible.
<b>2. In pregnancies with an increased risk of fetal CHD (see appendix A)</b>	<ul style="list-style-type: none"> <li>i. 18-21 weeks in majority of cases</li> <li>ii. Earlier in selected cases e.g. family history or increased nuchal translucency</li> <li>iii. Whenever referred if later than 20 weeks</li> </ul>
<b><i>Where should the scans be performed?</i></b>	
In all cases there should be essential service links outlined in basic requirements	<ul style="list-style-type: none"> <li>i. In a fetal cardiology unit</li> <li>ii. In a feto-maternal medicine unit</li> <li>iii. In a dedicated area and at a dedicated time in a paediatric cardiology unit, but not running concurrently with a paediatric cardiology clinic</li> </ul>
<b><i>Who should be scanned?</i></b>	
<b>1. Pregnancies at increased risk for fetal CHD (see appendix A)</b>	Pregnancies at increased risk for fetal CHD are commonly referred to tertiary centres for fetal cardiology assessment. The risk of having CHD depends on the individual high risk group, but the majority of these cases will be normal, with approximately 10-15% having CHD.
	The future aim for these pregnancies is that as obstetric screening improves the fetal heart in these cases could be examined in detail at the local hospital, with referral to the tertiary centre if normality cannot be confirmed or an abnormality is detected. Such development is much more likely if each obstetric ultrasound unit works towards having a local champion, who has appropriate training and experience to examine the fetal heart in detail and can decide between normality and abnormality, with appropriate support from a tertiary centre where needed
<b>2. Low risk pregnancies</b>	The majority of cases of fetal CHD occur in low risk pregnancies. These will only be detected during screening of low risk pregnancies by examining the fetal heart at the time of the obstetric anomaly scan.
	Standards for screening for heart defects during the routine 18+0 to 20+6 obstetric fetal anomaly scans have been recommended by NICE and RCOG and have been refined by FASP for national implementation (1,2,3). The BCCA supports these national guidelines and recommends the use of five views for cardiac evaluation during the fetal anomaly scan <ul style="list-style-type: none"> <li>i. Abdominal situs view</li> <li>ii. Four chamber view</li> <li>iii. View of aorta from left ventricle</li> <li>iv. View of pulmonary artery from right ventricle</li> <li>v. Three vessel / tracheal view</li> </ul>
	The referral pathway to the fetal cardiology service from the regional obstetric scanning services should be clearly defined.

## Counselling for prenatal diagnosis of congenital heart disease

<i>Information and Counselling</i>	
<b>1. Before the scan</b>	<ul style="list-style-type: none"> <li>i. Provide information leaflets</li> <li>ii. Make sure parents have understood the reason for referral for fetal cardiac evaluation</li> <li>iii. Explain implications of scan               <ul style="list-style-type: none"> <li>- Many pregnant women undergoing a routine ultrasound examination have not fully understood the implications of the scan, in particular that an abnormality in their baby may be revealed</li> </ul> </li> <li>iv. Explain what can and cannot be detected and the limitations of the scan</li> <li>v. Ensure parents want to know if there is a problem</li> <li>vi. Information must be provided in a format that is easy to understand and accessible to pregnant women with additional needs such as physical, sensory or learning impairments and for those who do not read or speak English</li> </ul>
<b>2. After the diagnosis of an abnormality</b>	<p>Following the detection of a problem, it is vital to be able to provide appropriate and adequate information, counselling and support. This should be done in a non-directive manner. The parents should be made aware of all options available to them and sufficient information and support must be provided to enable them to make the right decision for their individual circumstances.</p>
a) Who should counsel for fetal heart abnormalities?	<ul style="list-style-type: none"> <li>i. A fetal cardiologist or paediatric cardiologist with experience of fetal congenital heart disease, its associations and outcome, is the most appropriate person</li> <li>ii. There should be a nurse practitioner / counsellor /specialist practitioner present at initial discussions with parents and also in all follow-up discussions</li> <li>iii. Counselling can be done in conjunction with an obstetrician/fetal medicine expert/ geneticist/neonatologist/ or a paediatric cardiac surgeon where appropriate</li> </ul>
b) Where should the counselling take place?	In a quiet room separate from the scan room.

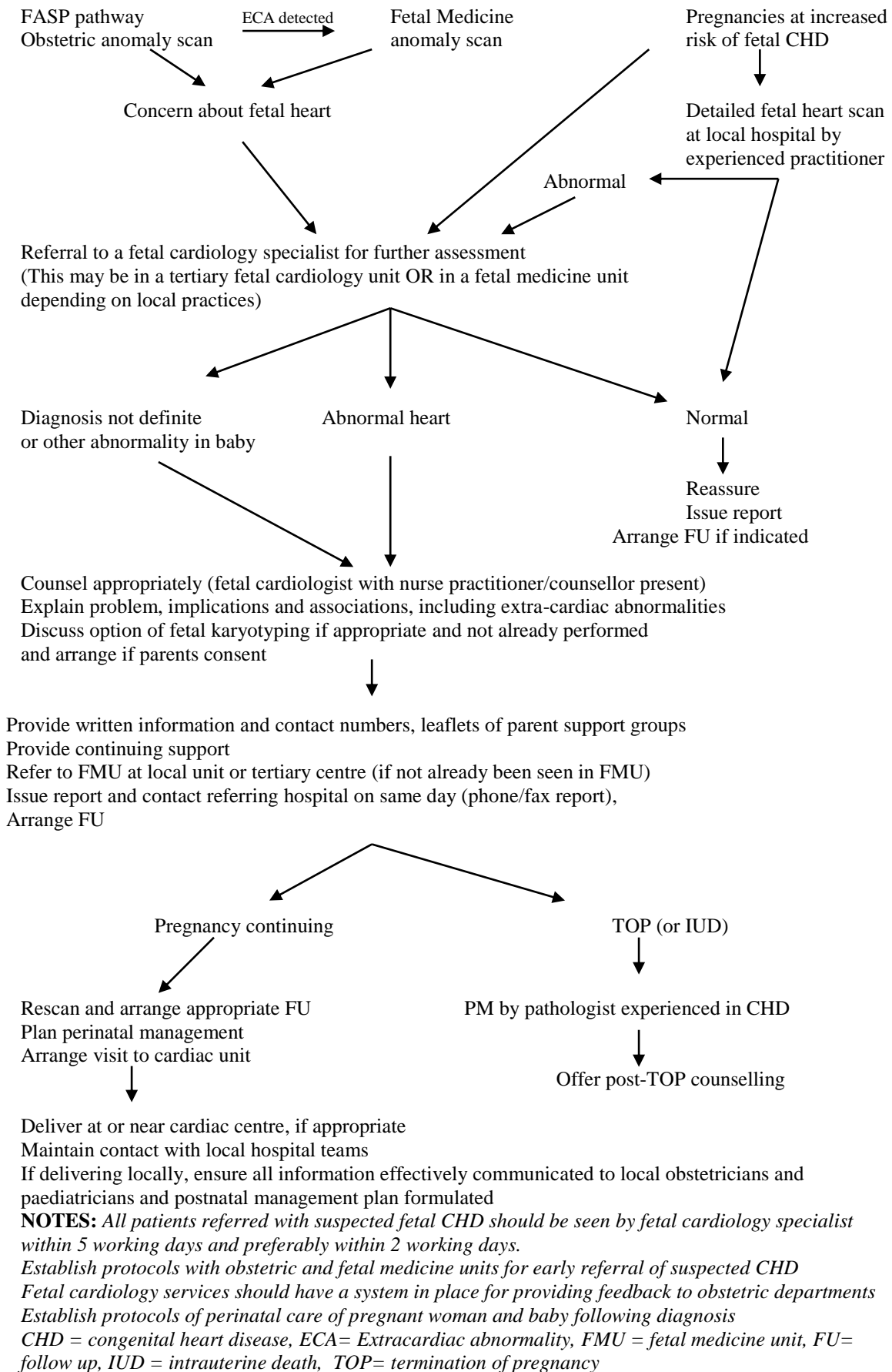
<p>c) Information to parents</p>	<ul style="list-style-type: none"><li>i. The clinician should be able to provide the parents with detailed information about their baby's heart problem, including an accurate description of the anomaly, information regarding the need for non-surgical or surgical intervention, the type of surgery available for the condition, the number of procedures likely to be required, the associated mortality and morbidity, and the overall long term outlook for the child. They should also explain all management options available, including termination of pregnancy where appropriate.</li><li>ii. Allow parents time for questions and to express grief and to 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 will make it difficult for them understand all the information in one quick sitting. No matter how well the initial explanation is made, reinforcement of the facts is likely to be required. A named nurse practitioner/ counsellor/specialist practitioner who has been present during the counselling session can provide reinforcement of the facts.</li><li>iii. Give parents written information to take away including information and contact numbers for relevant parent support groups.</li><li>iv. Give parents contact names and numbers of staff within unit and named nurse practitioner/ counsellor/specialist practitioner who can provide continuing support.</li><li>v. Make appropriate follow up arrangements.</li><li>vi. Refer to feto-maternal medicine unit in cases where pregnant lady has not already been assessed by fetal medicine specialist:<ul style="list-style-type: none"><li>a) exclude or define extent of any extra-cardiac malformations</li><li>b) discuss and consider fetal karyotyping where appropriate</li></ul></li><li>vii. Document all the discussions at the counselling session:<ul style="list-style-type: none"><li>a) consultant record</li><li>b) nurse practitioner/counsellor/specialist practitioner record</li></ul></li></ul>
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<i>Communication following initial diagnosis</i>	
	Must have defined mechanism of communication with relevant personnel. Pathways of communication with all those involved in the pregnant woman's care must be well defined. A copy of all reports should be placed in the pregnant woman's hand held obstetric notes and be available to anyone reviewing the lady.
<b>1. At specialist centre</b>	<ul style="list-style-type: none"> <li>i. Feto-maternal medicine unit (where relevant)</li> <li>ii. Genetics department (where relevant)</li> <li>iii. Others involved in care (where relevant)</li> </ul>
<b>2. At pregnant woman's local hospital</b>	<ul style="list-style-type: none"> <li>i. Obstetric consultant</li> <li>ii. Feto-maternal medicine unit (where present)</li> <li>ii. Liaison midwife</li> <li>iii. Feedback to obstetric ultrasound department</li> <li>iv. Local paediatrician (where relevant)</li> </ul>
<b>3. Other</b>	<ul style="list-style-type: none"> <li>i. General Practitioner</li> <li>ii. Community midwife (where relevant)</li> </ul>
<i>Management following initial diagnosis and counselling</i>	
	The parents may elect to stop the pregnancy or may decide on active treatment of the CHD for their baby. In both circumstances, they will need continuing support and provision must be made for this.
<b>1. In continuing pregnancies</b>	<ul style="list-style-type: none"> <li>i. Counsel after each subsequent scan.</li> <li>ii. Make appropriate arrangements for delivery: <ul style="list-style-type: none"> <li>a) deliver at or near cardiac centre if appropriate, with relevant multidisciplinary teams fully informed about mother and baby</li> <li>b) if a local delivery ensure the local obstetricians and paediatricians have relevant information about mother and baby and that a postnatal management plan is formulated for referral of baby to cardiac unit</li> </ul> </li> <li>iii. Make appropriate appointments for parents to see other personnel e.g. paediatric cardiac surgeon, neonatologist, paediatrician, geneticist, or to speak with other parents.</li> </ul>
<b>2. In difficult cases</b>	<ul style="list-style-type: none"> <li>i. Discuss with colleagues internally or externally.</li> <li>ii. Establish local and national network to facilitate this.</li> <li>iii. Refer for second opinion if requested by parents.</li> </ul>



<b>3. In cases of termination of pregnancy or intrauterine death</b>	<ul style="list-style-type: none"><li>i. Request autopsy in appropriate cases</li><li>ii. Autopsy, if performed, must be conducted by pathologist experienced in CHD</li><li>iii. Correlate echocardiogram and autopsy findings</li><li>iv. If no autopsy is performed, ensure digital clips of scan is stored, which can be validated by an experienced colleague if necessary</li></ul>
<b>4. Post termination/ pregnancy loss counselling</b>	<p>Following termination or spontaneous intrauterine loss, women and their partners should be offered further counselling.</p> <ul style="list-style-type: none"><li>i. Usually approximately 6 weeks later</li><li>ii. This may be provided by local obstetrician, tertiary specialists or a combination of both</li><li>iii. Ensure relevant and appropriate follow-up made</li></ul>

**PATHWAY FOR FETAL DIAGNOSIS OF CONGENITAL HEART DISEASE**



## **Summary of Minimum Fetal Cardiology Service Standards**

<p><u>Staffing</u></p> <ul style="list-style-type: none"><li>• Consultant(s) trained in fetal cardiology</li><li>• Specialist practitioners to provide support for parents</li></ul>
<p><u>Setting</u></p> <ul style="list-style-type: none"><li>• Dedicated fetal cardiology clinic(s)</li><li>• Adequate time for scanning and counselling (minimum 45 minutes)</li></ul>
<p><u>Equipment</u></p> <ul style="list-style-type: none"><li>• High resolution scanner</li><li>• Database for data entry and reports</li><li>• Image archive system</li></ul>
<p><u>Safety</u></p> <ul style="list-style-type: none"><li>• As per NHS FASP recommendations and guidelines</li></ul>
<p><u>Access</u></p> <ul style="list-style-type: none"><li>• Rapid access appointments for suspected abnormalities (Definitely within 5 working days but preferably within 2 working days)</li><li>• At optimum time (18-21 weeks gestation or earlier) for pregnancies at increased risk for fetal CHD</li></ul>
<p><u>Consultation</u></p> <ul style="list-style-type: none"><li>• Accurate and detailed diagnosis</li><li>• Explanation of findings, implications, management and prognosis to parents</li><li>• Discussion of risk of associated anomaly and option of fetal anomaly scan and karyotyping where relevant</li><li>• Discussion of all management options available</li><li>• Provision of written information</li></ul>
<p><u>On-going care</u></p> <ul style="list-style-type: none"><li>• Direct access to specialist practitioners for support and information</li><li>• Parental contact with services to be involved in care after birth</li><li>• Follow up fetal cardiac consultations</li></ul>
<p><u>Effective networks</u></p> <ul style="list-style-type: none"><li>• Sonographers and Obstetricians for streamlined referrals, training and feedback</li><li>• Feto-maternal Medicine Units for assessment whole baby and karyotyping</li><li>• Obstetric services for delivery planning</li><li>• Neonatal and paediatric services for management at birth</li><li>• Paediatric cardiac services</li></ul>
<p><u>Audit</u></p> <ul style="list-style-type: none"><li>• Detection rate</li><li>• Diagnostic accuracy</li><li>• Outcome</li></ul>

## **Appendix A - Pregnancies at increased risk for fetal CHD, including functional and rhythm disturbances where referral for detailed fetal echocardiography should be considered**

### **Maternal indications**

- 1) Maternal congenital heart disease (risk 2-6%)
- 2) Maternal metabolic disorders, especially if poor control in early gestation
  - i. diabetes mellitus (risk 2-3%)
  - ii. phenylketonuria (risk 8-10%)
- 3) Maternal exposure to cardiac teratogens:
  - i. anticonvulsant, retinoic acid, lithium (risk 2%)
  - ii. viral infection (rubella, CMV, coxsackie, parvovirus) and toxoplasma
- 4) Maternal collagen disease with anti Ro/SSA and/or anti La/SSB (Risk 2-3%)
- 5) Maternal medication with NSAID drugs

### **Familial indications**

- 1) Paternal congenital heart disease (risk 2-6%)
- 2) Previous child or fetus with congenital heart disease or congenital heart block
  - a. 1 affected child (risk 2-3%, though higher for some lesions, for example, isomerism)
  - b. 2 affected children (risk 10%)
  - c. 3 affected children (risk 50%)
- 3) Previous child with congenital complete heart block with maternal auto antibodies (risk 20% for CHB)
- 4) Chromosomal anomalies, gene disorders or syndromes with congenital heart disease or cardiomyopathy ( risk will depend on individual disorder)

### **Fetal indications**

- 1) Suspicion of fetal cardiac abnormality during an obstetric scan**
  - i. Most cases of cases of fetal congenital heart disease will occur in this group
- 2) Fetal arrhythmias
  - i. sustained bradycardia heart rate <120 beats per minute
  - ii. tachycardia – heart rate >180 beats per minute

(Irregular heart rhythms can be managed in conjunction with the local obstetric teams. In many cases referral to tertiary centre can be avoided if agreed management protocols are in place locally.)
- 3) Nuchal translucency >99<sup>th</sup> centile for crown rump length (= or >3.5mm)
  - i. Risk 6-7% for NT >3.5 mm but increases with increasing NT measurement
  - ii. A nuchal translucency >95<sup>th</sup> centile is also associated with an increased risk of CHD but with lower risk and due to the workload

involved, local policies will determine whether this group should be offered a detailed cardiac scan

- 4) Fetal hydrops
- 5) Pericardial effusion
- 6) Pleural effusion
- 7) Polyhydramnios
- 8) Extra-cardiac malformation
- 9) Chromosomal abnormalities
- 10) Genetic syndromes
- 11) Monochorionic twins (risk 8-10%)
  
- 12) Other states with known risk for fetal heart failure:
  - i. tumors with a large vascular supply
  - ii. arteriovenous fistulas
  - iii. absence of ductus venosus
  - iv. acardiac twin
  - v. twin-twin transfusion syndrome
  - vi. fetal anaemia

**Note: Initial assessment of many of the above cases could be made by a fetal medicine specialist or by experienced sonographer who have had appropriate training in fetal heart scanning. Those with a suspected cardiac abnormality can then be referred to a fetal cardiology specialist for further assessment.**

**The original version of these standards was produced following consultation with the membership of the BCCA and approval from BCCA council in March 2010.**

**The standards were revised in April 2012 (G Sharland) following discussions and consultation with the NHS Fetal Anomaly Screening Programme team and consultation with BCCA membership.**

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