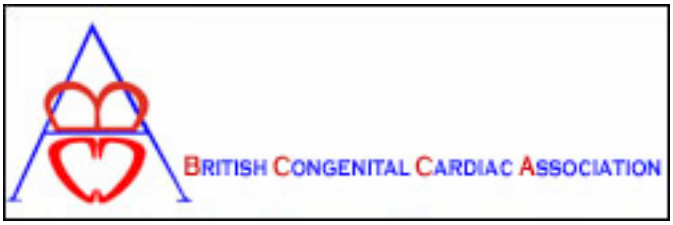

Fetal Cardiology Standards

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

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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. 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. The means of achieving a more uniform national standard is under review (3).

The standards 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 services, which can be adapted to fit in with local models of delivery.

Standards for fetal cardiology service in a tertiary centre

Aims and role of a fetal cardiology service
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 on the cardiac scan that suggest there may be an extra-cardiac abnormality, even though the heart structure is normal. 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 management of on-going pregnancy in collaboration with the pregnant woman's obstetrician, and all personnel likely to be involved in perinatal management, in order to try and improve outcome.
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.

	Basic requirements for a fetal cardiology service
1. STAFFING	Dedicated multidisciplinary team trained in the diagnosis and management of fetal CHD and related abnormalities.
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 trained in fetal cardiology 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	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.
c) Other clinical staff	The following can perform scans under supervision of a consultant trained in fetal cardiology: i. Sonographers

	ii. Specialist radiographers iii. Nurse practitioners iv. Doctors in training
2. TIME FOR SCANS	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.
3. RELATED SERVICES a) Essential	There must be well established links with the following services: 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
4. EQUIPMENT	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
5. DATABASE AND IMAGE STORAGE	A record must be kept of all scans performed. i. Must have database for data entry on all scans ii. Must have system of obtaining and recording outcomes for audit iii. Aim for national database for all fetal cardiac diagnosis iv. Must keep videotape/digital recording of all scans

	At what stage should scans be performed?
1) In case of a suspected cardiac problem	Preferably within 2 working days of referral but definitely within one week
2) In high risk groups	i. 18-21 weeks in majority of cases ii. Earlier in selected cases e.g. family history or increased nuchal translucency iii. Whenever referred if later than 20 weeks

	Where should scans be performed?
In all cases should have essential service links outlined in basic requirements	i. In the fetal cardiology unit ii. In the feto-maternal medicine unit iii. In a dedicated area and at a dedicated time in a paediatric cardiology unit, but not running concurrently with a paediatric cardiology clinic

	Who should be scanned?
1. HIGH RISK PREGNANCIES	High risk pregnancies as outlined in appendix A are commonly referred to tertiary centres for fetal cardiology assessment. The risk of having CHD depends on the individual high risk group. However the majority of these cases will be normal with approximately 10% having CHD.
	As obstetric screening improves it may be possible to check the fetal heart in these cases at the local hospital, with referral to the tertiary centre if normality cannot be confirmed. Such development is much more likely if each obstetric ultrasound unit has a local champion with appropriate training and experience to examine the fetal heart in detail and decide between normality and abnormality, with appropriate support from a tertiary centre where needed (This is in accordance with FASP recommendations (3)).
2. LOW RISK PREGNANCIES	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 obstetric 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 their recommended views for cardiac evaluation to be included in the fetal anomaly scan.
	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

	Information and Counselling
1. BEFORE THE SCAN	i. Provide information leaflets ii. Make sure parents have understood the reason for referral for fetal cardiac evaluation iii. Explain implications of scan - 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 iv. Explain what can and cannot be detected and the limitations of the scan v. Ensure parents want to know if there is a problem
2. AFTER THE DIAGNOSIS OF AN ABNORMALITY	Following the detection of a problem, it is vital to be able to provide appropriate and adequate information, counselling and support.
a) Who should counsel for fetal heart abnormalities?	i. A fetal cardiologist or paediatric cardiologist with experience of fetal congenital heart disease, its associations and outcome, is the most appropriate person ii. There should be a nurse practitioner / counsellor /specialist practitioner present at initial discussions with parents and also in all follow-up discussions iii. Counselling can be done in conjunction with an obstetrician/fetal medicine expert/ geneticist/neonatologist/ or a paediatric cardiac surgeon where appropriate
b) Where should the counselling be done?	In a quiet room separate from the scan room.

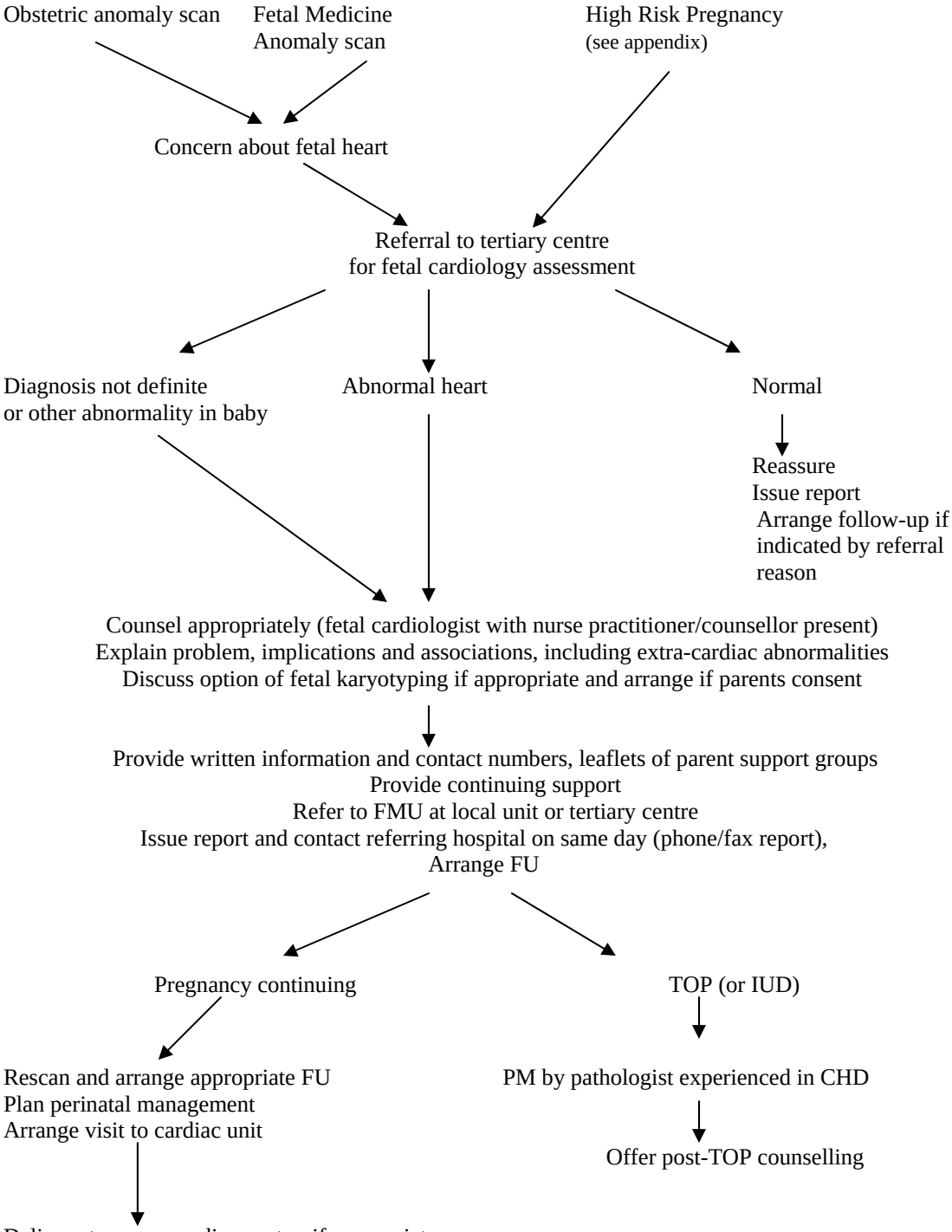
c) Information to parents	i.	The clinician should be able to provide the parents with detailed information about their baby's heart problem, including an accurate description of the anomaly, information regarding the need for non-surgical or surgical intervention, the type of surgery available for the 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, including termination of pregnancy where appropriate.
	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.
	iii.	Give parents written information to take away including information and contact numbers for relevant parent support groups.
	iv.	Give parents contact names and numbers of staff within unit and named nurse practitioner/ counsellor/specialist practitioner who can provide continuing support.
	v.	Make appropriate follow up arrangements.
	vi.	Refer to feto-maternal medicine unit: a) exclude or define extent of any extra-cardiac malformations b) discuss and consider fetal karyotyping where appropriate
	vii.	Document all the discussions at the counselling session: a) consultant record b) nurse practitioner/counsellor/specialist practitioner record

	Communication following initial diagnosis
	Must have defined mechanism of communication with relevant personnel.
At specialist centre	i. Feto-maternal medicine unit (where relevant) ii. Genetics department (where relevant) iii Others involved in care (where relevant)
At pregnant woman's local hospital	i. Obstetric consultant ii Feto-maternal medicine unit (where present) ii Liaison midwife iii. Feedback to obstetric ultrasound department iv. Local paediatrician (where relevant)
Other	i. General Practitioner

	Management following initial diagnosis and counselling
	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.
1. In continuing pregnancies	i. Counsel after each subsequent scan. ii. Make appropriate arrangements for delivery: a) deliver at or near cardiac centre if appropriate, with relevant multidisciplinary teams fully informed about mother and baby 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 iii. Make appropriate appointments for parents to see other personnel e.g. paediatric cardiac surgeon, neonatologist, paediatrician, geneticist, or to speak with other parents.
2. In difficult cases	i. Discuss with colleagues internally or externally. ii. Establish local and national network to facilitate this. iii. Refer for second opinion if requested by parents.
3. In cases of termination of pregnancy or intrauterine death	i. Request autopsy in appropriate cases ii. Autopsy, if performed, must be conducted by pathologist experienced in CHD iii. Correlate echocardiogram and autopsy findings iv. If no autopsy is performed, ensure video recording of scan is stored, which can be validated by an experienced colleague if necessary

4. Post termination/ pregnancy loss counselling	Offer in all cases following termination or spontaneous intrauterine loss: i. Usually approximately 6 weeks later ii. Where appropriate, may be done with obstetric team iii. Ensure relevant and appropriate follow-up made

PATHWAY FOR FETAL DIAGNOSIS OF CONGENITAL HEART DISEASE



NOTES: *All patients referred with suspected fetal CHD should be seen within 1 week and preferably within 2 working days.*
Establish protocols with obstetric and fetal medicine units for early referral of suspected CHD
The fetal cardiology service to have a system in place for providing feedback to obstetric departments
Establish protocols of perinatal care of pregnant woman and baby following diagnosis

Summary of Minimum Fetal Cardiology Service Standards

Staffing • Consultant(s) trained in fetal cardiology • Specialist practitioners to provide support for parents
Setting • Dedicated fetal cardiology clinic(s) • Adequate time for scanning and counselling (minimum 45 minutes)
Equipment • High resolution scanner • Database for data entry and reports • Image archive system
Access • Rapid access appointments for suspected abnormalities (Definitely within 1 week but preferably within 48 hours) • At optimum time (18-21 weeks gestation or earlier) for high risk pregnancies
Fetal Cardiology Consultation • Accurate and detailed diagnosis • Explanation of findings, implications, management and prognosis to parents • Discussion of risk of associated anomaly and option of fetal anomaly scan and karyotyping if relevant • Discussion of option of termination for major abnormalities • Provision of written information
On-going care • Direct access to specialist practitioners for support and information • Parental contact with services to be involved in care after birth • Follow up fetal cardiac consultations
Effective networks • Sonographers and Obstetricians for streamlined referrals, training and feed-back • Feto-maternal Medicine Units for assessment whole baby and karyotyping • Obstetric services for delivery planning • Neonatal and paediatric services for management at birth • Paediatric cardiac services
Audit • Detection rate • Diagnostic accuracy • Outcome

Appendix A - **Referral indications for fetal echocardiography**

Maternal indications

1) Maternal congenital heart disease
2) Maternal metabolic disorders, especially if poor control in early gestation
    i. diabetes mellitus
    ii. phenylketonuria

3) Maternal exposure to cardiac teratogens:
    i. anticonvulsant, retinoic acid, lithium
    ii. viral infection (rubella, CMV, coxsackie, parvovirus) and toxoplasma
4) Maternal collagen disease with anti Ro/SSA and/or anti La/SSB
5) Maternal medication with NSAID drugs after 25-30 gestational weeks

Familial indications

1) Paternal congenital heart disease
2) Previous child or fetus with congenital heart disease or congenital heart block
3) Chromosomal anomalies, gene disorders or syndromes with congenital heart disease or cardiomyopathy

Fetal indications

1) Suspicion of fetal cardiac abnormality during an obstetric scan
2) Fetal hydrops
3) Pericardial effusion
4) Pleural effusion
5) Polyhydramnios
6) Extra-cardiac malformation
7) Chromosomal abnormalities
8) Genetic syndromes
9) Nuchal translucency >99th centile for crown rump length (>3.5mm)
    (A nuchal translucency >95th centile is also associated with an increased risk of CHD but due to the workload involved, local policies will determine whether this group should be offered a detailed cardiac scan)
10) Monochorionic twins
11) 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.)
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

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

References

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2) National Institute for Clinical Excellence Antenatal care – routine care for the healthy pregnant woman Clinical Guideline March 2008 www.nice.org.uk

3) UK National Screening Committee – Fetal Anomaly Screening Programme (FASP) www.fetalanomaly.screening.nhs.uk

4) Paediatric cardiology curriculum and sub-specialty training. www.jrcptb.org.uk

5) Recommendations from the Association for European Paediatric Cardiology for training in Fetal Cardiology, 2008. www.aepc.org/aepc/nid/Fetal%20Cardiology