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Recommendations of the AEPC

Recommendations for the practice of fetal cardiology in Europe

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Developed by the Fetal Cardiology Working Group of the Association for European Paediatric Cardiology

FETAL CARDIOLOGY IS CURRENTLY PRACTISED IN most European countries, but even within countries there is a great variation in the service provided. The recommendations provided in this document are intended to be guide for all paediatric cardiologists undertaking fetal echocardiography with the view of providing a service in fetal cardiology. It is clear that the health and legal systems vary from country to country, so that not all aspects of these recommendations can be implemented in all countries. The recommendations, nonetheless, provide a framework that can be adapted to fit in with local situations.

Whilst we are aware that, in some countries, our obstetric colleagues perform a significant proportion of fetal echocardiograms, these recommendations are designed specifically for the paediatric cardiologist. Although we have included a section on cardiac evaluation during obstetric scans, and have made proposals for sonographers and obstetricians, we would greatly support the formulation of parallel recommendations for obstetricians. The fetal cardiology working group is very keen to help develop these in collaboration with a society such as The International Society of Ultrasound in Obstetrics and Gynecology.

How to organise a service for fetal cardiology

1. Aims of service

a. To make an accurate diagnosis of cardiac abnormalities, if present in the fetus, as early as possible in gestation. This includes the diagnosis of structural defects, functional defects, and disturbances of rhythm. An early accurate diagnosis will make possible parental choice, as well as providing the opportunity to plan the delivery and postnatal management so as to optimize the outcome. Support can also be provided to specialists in fetal medicine, and to obstetricians, in the management of fetuses with functional disturbances, as in twin-to-twin transfusion syndrome
b. To provide appropriate counselling and support for parents and families following prenatal diagnosis
c. To communicate results to the referring obstetrician or general practitioner
d. To plan management of on-going pregnancies in collaboration with the obstetrician, and all personnel likely to be involved in peri-natal management.
e. To initiate prenatal treatment where appropriate, such as in fetal arrhythmias and selected structural lesions
f. To maintain a database to enable regular audit of activity, and to obtain data concerning outcomes. The data should include information on the monitoring of sensitivity and specificity, and the incidence of false negative and false positive diagnoses.

2. Basic requirements

a. Space

i. In the Department of Paediatric Cardiology
ii. In the unit for fetal medicine or obstetrics
iii. Other
iv. Ideally the service should have links to the unit providing services in paediatric cardiology and paediatric cardiothoracic surgery, and to the units concerned with fetal medicine and obstetrics. It is beneficial also to have links to the genetics department and the service for adult cardiology.

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b. Equipment: High resolution equipment will allow earlier and more accurate diagnosis and also quicker evaluation
   i. Similar to requirements for paediatric echocardiography
   ii. 3.5, 5.0 and 7.5 MHz transducers
   iii. The availability of an obstetric probe is beneficial
   iv. M-mode, pulsed wave and continuous wave Doppler and colour flow
   v. Transvaginal probes may be a desirable addition if very early scans are undertaken. They are more likely to be used, however, in a fetal medicine unit where appropriately trained staff and more private scanning conditions are available.

c. Database
d. Facilities for the storage of videotape or digital recording of all scans at least until the outcome has been confirmed, preferably much longer depending on the legal requirements in each country.

3. Staff

a. Medical staff
   i. Staff at consultant grade, and trained in fetal echocardiography, must be available to check scans as necessary, and oversee all problems.

b. Other staff who scan under supervision of consultant
   i. Sonographers
   ii. Nurse practitioners
   iii. Doctors in training.

c. Administrative staff to make appointments and help collect data
d. A nurse/counsellor to provide help and support to families.

4. How many scans need to be performed to maintain a service at an adequate level within the unit?

a. 250–500 normal studies each year
b. Approximately 50 abnormalities each year.

5. At what stage should scans be performed?

a. The earlier the better – but all early scans should have a further scan between the 18th and 20th weeks of gestation. If a termination is discussed because of an early diagnosis, and a repeat scan therefore is not possible, peer review of the findings is desirable
b. Between the 13th and 14th weeks in selected cases
c. Between the 18th and 20th weeks in the majority
d. Whenever referred, if later than 20 weeks.

Counselling for prenatal diagnosis of cardiac disease

1. Before the scan
   a. Make sure parents have understood why they have been referred for the scan
   b. Explain the implications of the scan
   c. Explain what can and cannot be detected
   d. Ensure parents want to know if there is a problem.

2. After diagnosis of a problem
   a. Move parents out of the scanning room into a separate room for counselling
   b. Counsel parents in the presence of the counsellor or nurse practitioner, or some other medical person
   c. Document the proceeding of the counselling session.

3. Who should counsel parents for fetal congenital cardiac disease?
   a. A paediatric or fetal cardiologist must be involved
   b. Counselling can be done in conjunction with the obstetrician, or an expert in fetal medicine, or a geneticist, neonatologist, or paediatric cardiac surgeon, but remember that it is inappropriate and insensitive to have too many people in the room
   c. It is beneficial to have the involvement of a nurse, counsellor, or midwife.

4. After giving parents information about the cardiac problem
   a. Allow parents time for questions, to express grief, and to be left alone if desired
   b. Give parents written information to take away, including contact numbers and information about relevant support groups
   c. Make appropriate arrangements for follow-up
   d. Refer to the fetal medicine unit for consideration of karyotyping
   e. Communicate with the local hospital and general practitioner.

5. In cases of termination of pregnancy
   a. Autopsy
      i. Correlate the findings at autopsy with those from the echocardiographic study
      ii. In case no autopsy has taken place, ensure that a video recording of the scan is retained, which can be validated by an experienced colleague if necessary.
   b. Post-termination counselling.

6. *In continuing pregnancies*
   a. Counsel after each subsequent scan
   b. Make appropriate arrangements for delivery
   c. Make appropriate appointments for the parents to see other personnel, such as the paediatric cardiac surgeon, neonataologist, geneticist, or to speak with other parents.

Indications for referral for fetal echocardiography

1. *Maternal indications*

a. Maternal metabolic disorders, especially if poor control in early gestation
   i. Diabetes mellitus
   ii. Phenylketonuria.
b. Maternal exposure to cardiac teratogens
   i. Anticonvulsants, retinoic acid, lithium
   ii. Viral and other infections, such as rubella, cytomegalovirus, coxsackie, parvo, and toxoplasma.
c. Maternal collagen disease with anti Ro/SSA and/or anti La/SSB antibodies
d. Maternal congenital cardiac disease and familial cardiomyopathy
e. Maternal medication with nonsteroidal anti-inflammatory drugs after 25 to 30 gestational weeks.

2. *Familial indications*

a. Paternal congenital cardiac disease
b. Previous child or fetus with congenital cardiac disease or congenital heart block
c. Chromosomal anomalies, gene disorders, or syndromes with congenital cardiac disease or cardiomyopathy.

3. *Fetal indications*

a. Suspicion of cardiac malformation or disease during an obstetrical scan
b. Fetal hydrops
c. Hydrothorax
d. Polyhydramnios
e. Extracardiac malformation, especially:
   i. Omphalocele
   ii. Diaphragmatic hernia
   iii. Duodenal atresia
   iv. Tracheo-esophagal fistula
   v. Cystic hygroma.
f. Chromosomal abnormalities
g. Nuchal translucency greater than the 99th centile for crown rump length
h. Arrhythmias
   i. Sustained bradycardia of less than 100 beats/min
   ii. Tachycardia, either intermittent or sustained, if greater than 180–200 beats/min
   iii. Frequent ectopic beats.
i. Other states with a known risk for fetal heart failure:
   i. Tumours with a large vascular supply
   ii. Arteriovenous fistulas
   iii. Absence of the venous duct ("ductus venosus")
   iv. Acardiac twin
   v. Feto-fetal transfusion syndrome
   vi. Anaemia.
j. Monochorionic twins because of the associated increased risk of fetal structural cardiac malformations.

Notes on the indications for referral

The recommendations for referral can be implemented in different ways and to different degrees, depending on local practices and the workload of individual centers. Many obstetricians working in fetal medicine are very skilled in making both structural and functional diagnosis of the heart. Thus, in some countries, functional studies in intrauterine growth retardation, or the twin-twin transfusion syndrome, are predominantly made by obstetricians. Thus, collaboration must take place between obstetricians and paediatric cardiologists in the management of such cases.

What comprises a complete fetal echocardiogram

A. *Recommendations for the fetal cardiologist offering a referral service for fetal echocardiography for mothers at increased risk for fetal cardiac anomalies*

1. Normal arrangement of the aorta and inferior caval vein in the abdomen
2. Stomach and heart on the left side of the fetus
3. A normal four chamber view
   a. Size
      i. About one third of the thorax
         1. Any suspicion of abnormal size should prompt measurement of cardiothoracic ratio for comparison with normal range.
   b. Position
      i. Septum at an angle of about 45° to the midline.
   c. Structure
      i. Two atriums of approximately equal size
      ii. Ventricular morphology
      iii. Two ventricles having cavities of approximately equal size and mural thickness, and remembering that scans after 30 weeks do not necessarily show equal sized ventricles

1. Any suspicion of abnormal size should prompt measurement for comparison with normal ranges.

iv. Two atrioventricular valves opening during diastole
v. Intact crux of the heart, with off-set hingepoints of the atrioventricular valves
vi. Intact ventricular septum from apex to crux and from crux to the origin of the aorta
vii. Defect in the oval foramen ("foramen ovale") in the middle third of the atrial septum, with the flap valve in the left atrium
viii. Pulmonary vein(s) to left atrium, with at least one vein visualized by colour flow mapping
ix. Systemic veins connecting to the right atrium.

d. Function
i. Equally well contracting ventricles.

4. Aortic origin from the left ventricle, with the septum continuous with the anterior wall of the aorta
5. Pulmonary trunk arising from the right ventricle
6. Pulmonary trunk slightly bigger than the aorta at 16 to 20 weeks gestation
   a. Any suspicion of abnormal size should prompt measurement for comparison with normal ranges
7. Pulmonary trunk crossing over the aortic origin
8. At least one pulmonary artery of normal size
9. Arterial duct lying caudal to the transverse arch
10. Duct and arch of approximately equal size
    a. Any suspicion of abnormal size should prompt measurement for comparison with normal ranges
11. Head and neck vessels arising from the arch
12. Unaliased colour flow in the appropriate direction across all four valves
    a. If aliasing occurs at any point, pulsed Doppler should be used to obtain an exact velocity
13. Unaliased colour flow in the appropriate direction in the duct and aortic arch
    a. If aliasing, pulsed Doppler should be used to obtain an exact velocity
14. No colour flow crossing the ventricular septum
15. No regurgitation evident at any valve on colour flow mapping
16. Normal heart rate and rhythm
    a. If rate or rhythm are abnormal, M-mode or pulsed Doppler should be used to demonstrate the relationship between atrial and ventricular contraction.

B. Recommendations for cardiac evaluation during every obstetric ultrasound scan

Although views visualising the outflow tracts are not included in this section, they are important, and anybody performing obstetric cardiac screening should have the ambition to move to the level recommended in section C as soon as possible.

1. Stomach and heart on the left side of the fetus
2. A normal four chamber view
   a. Size
      i. About one third of the thorax.
   b. Position
      i. Septum at an angle of about 45° to the midline.
   c. Structure
      i. Two atriums of approximately equal size
      ii. Two ventricles having cavities of approximately equal size and mural thickness
      iii. Two opening atrioventricular valves
      iv. Intact crux of heart
      v. Intact ventricular septum from apex to crux
      vi. Defect in oval foramen in middle third of atrial septum.
   d. Function
      i. Equally contracting ventricles.

C. Recommendations for obstetricians, sonographers or radiologists who want to extend their evaluation of the heart, although not offering a specialist service

1. Stomach and heart on the left side of the fetus
2. A normal four chamber view
   a. Size
      i. About one third of the thorax.
   b. Position
      i. Septum at an angle of about 45° to the midline.
   c. Structure
      i. Two atriums of approximately equal size
      ii. Two ventricles having cavities of approximately equal size and mural thickness
      iii. Two atrioventricular valves opening during diastole
      iv. Intact crux of heart
      v. Intact ventricular septum from apex to crux
      vi. Defect in oval foramen in middle third of atrial septum.
   d. Function
      i. Equally contracting ventricles.
3. Aortic origin from left ventricle, with septum continuous with anterior wall of the aorta
4. Pulmonary trunk arising from right ventricle
5. Pulmonary trunk slightly bigger than aorta
6. Pulmonary trunk crossing over the aortic origin
7. Duct lying caudal to the transverse arch

8. Duct and arch of approximately equal size
9. Unaliased colour flow throughout the heart
10. Normal heart rate and rhythm.

Training in fetal cardiology

Background and specific training in fetal cardiology

Physicians wishing to be experts in fetal cardiology must be paediatric cardiologists. On the web page of the Association for European Paediatric Cardiology (www.AEPC.org), detailed instructions for training in order to become a paediatric cardiologist has already been published. All prospective fetal cardiologists should receive training at a recognized center for fetal cardiology under the guidance and supervision of experienced fetal cardiologist(s). The end result of training should be a fetal cardiologist capable of high diagnostic accuracy, who is able to give an informed prognosis, has the requisite skills in counselling, and who recognizes the limitations of his/her own knowledge and experience. He/she should also have demonstrated effective communication with and understanding of the role of other relevant subspecialists, such as in fetal medicine, obstetrics and clinical genetics.

There will be a natural variation in the ease with which individuals will achieve this goal. Giving guidance as to the number of procedures, therefore, serves as a recommendation only, and the aim should be to achieve the goals listed above.

Basic knowledge

1. General knowledge of pediatric cardiology
2. Understanding of the physical principles of ultrasound examination:
   a. Safety of ultrasound during fetal life
   b. Equipment required for the different kinds of examinations, and when the equipment can be used, especially concerning scans during the first trimester.
3. Obstetric scans are performed and interpreted by obstetricians, but fetal cardiologists should have basic knowledge about:
   a. How to evaluate number of fetuses, position and major fetal measurements
   b. Markers of chromosomal and genetic disorders
   c. Anomalies of other organs that can coexist with cardiac defects
   d. Characteristics of the fetal cardiovascular system
      i. Flow patterns in specific fetal vessels such as the venous and arterial ducts, the umbilical vessels, and the middle cerebral artery
      ii. Signs of fetal cardiac failure.
4. Basic knowledge in clinical genetics:
   a. Association between fetal cardiac defects and genetic syndromes
   b. When and how the karyotype can be obtained
   c. Molecular techniques in modern genetics, and its usefulness in fetal cardiology, such as in the evaluation of 22q11 microdeletion.

Teaching schedule

1. Performing fetal scans
   a. At a tertiary level in the department of perinatology or fetal cardiology
   b. At least 100 normal scans with evaluation of:
      i. Number of fetuses
      ii. Fetal position
      iii. Major fetal measurements, such as biparietal diameter, femoral length, occipitofrontal diameter, cranial and abdominal circumferences
      iv. Umbilical flow
      v. Basic evaluation of fetal organs. The fetal cardiologist is not responsible for the diagnosis of extracardiac abnormalities but should be familiar with the normal and abnormal ultrasonic appearances of major fetal organs.
2. All trainees in fetal cardiology should have demonstrated competence in demonstrating normal cardiac anatomy in at least 100 fetuses without other assistance than the routine checks of scans according to the policy of the individual unit
   a. Position of the heart
   b. Segmental analysis
   c. M-mode
   d. Color Doppler
   e. Pulsed Doppler
   f. Continuous wave Doppler, if necessary
   g. Other techniques, if deemed necessary
   h. Training should continue for the time needed by the supervising fetal cardiologist to confirm that the trainee has performed at least 100 normal fetal echoes without assistance.
3. Fetal echocardiography – abnormal scans
   a. Evaluation of fetal abnormalities. A minimum of 20 abnormalities should have been assessed under supervision
   b. Evaluation of fetal abnormalities off-line – A minimum of 50 cardiac abnormalities should have been evaluated from video recordings or other retrospective records. The trainee should have demonstrated diagnostic competence, documented by the supervising cardiologist
   c. Functional abnormalities – The trainee should gain experience in diagnosing arrhythmias as well as abnormal flow patterns in fetal vessels in the structurally normal heart, paying attention to interpretation and decision-making in the perinatal period.

4. Fetal cardiology courses
   a. One basic fetal echocardiography course
   b. Understanding of fetal cardiac pathology, either at a course or by attendance at a pathology department with experience in fetal cardiology.
5. Fetal therapy
   a. Arrhythmias – Good knowledge is required of the strategies for treatment or non-treatment of fetal arrhythmias. The trainee should be aware of the results of different therapies, and the potential adverse effects of drug treatment, including the effects on the mother, and should have been instructed in the strategies for monitoring treatment
   b. Invasive fetal therapy
      i. Indications, such as possibly some fetal arrhythmias, and structural lesions such as aortic valvar stenosis
      ii. Fetal cardiologists should be up to date with all possible fetal therapy, including experimental procedures. They must know the early and late results, as well as possible complications.
6. Issues of management
   a. Place and timing of delivery
   b. Planning of postnatal assessment and treatment, such as prostaglandin therapy and Rashkind septostomy.
7. Parental counselling
   This is a particularly sensitive area where the inexperienced trainee should be closely supervised.