



Tiny Tickers

Prenatal Congenital Heart Disease a new beginning



**Summary of issues and recommendations for improving
the prenatal detection, diagnosis and care of
Congenital Heart Disease in the UK**

FURTHER INFORMATION

ABOUT THIS DOCUMENT

This document proposes an integrated strategy for the prenatal (or antenatal) screening of babies for congenital heart disease (CoHD), and subsequent referral, diagnosis and management of the affected pregnancy.

We highlight current issues and recommend a care pathway (Map 1) and professional process (Map 2) to address gaps in training and provision of service.

If you have comments about this document, please contact us:

email: info@TinyTickers.org

FULL REPORT

This summary is based on the full report of the outcome of the Tiny Tickers RCOG Workshop, July 2008, on improving antenatal congenital heart disease, with 30 invited professionals. The full report, "Antenatal CoHD Pathway TTV1.pdf", including current practices, issues and recommendations, is available on the Tiny Tickers website:

www.tinytickers.org/infoprof_links.html (Link: "Antenatal CoHD Pathway TTV1.pdf")

ABOUT TINY TICKERS

Tiny Tickers is a national charity, established in 1999, working locally and nationally to improve the prenatal detection, diagnosis and care of congenital heart disease.

www.tinytickers.org

ANTENATAL DATA ONLINE

The Central Cardiac Audit database (CCAD) have begun to publish antenatal data derived from infants with major antenatal CoHD, by Strategic Health Authority:

www.ccad.org.uk/002/congenital.nsf/vwContent/Antenatal%20Diagnosis (accessed Feb. '10)

BINOCAR (British Isles Network of Congenital Anomaly Registers), collect data on congenital anomalies from various regional registers, where they exist: www.binocar.org

ANTENATAL CARDIAC TRAINING/CERTIFICATION RESOURCES

There are a number of online resources and CDs from organisations such as International Society of Ultrasound in Obstetrics and Gynaecology (ISUOG), Fetal Medicine Foundation (FMF) and Tiny Tickers. If you have information or comments about any of these resources, please contact us by email: info@TinyTickers.org

INTRODUCTION

Congenital Heart Disease is common

The heart is fully formed in the first 50 days of life.

Congenital heart disease (CoHD) is the most common congenital condition and accounts for 10% of all infant deaths in the developed world. Just under 1% of all pregnancies are affected by CoHD and usually there is no identifiable cause.

In the UK, with 800,000 births per annum (www.statistics.gov.uk), about 5,500 babies will be born with CoHD.

Half of these “heart babies” will have major CoHD, requiring surgery or intervention in the first year of life. One third of babies with major CoHD will also have duct-dependent heart disease and, if not recognised in the first few days after birth, this is life-threatening.

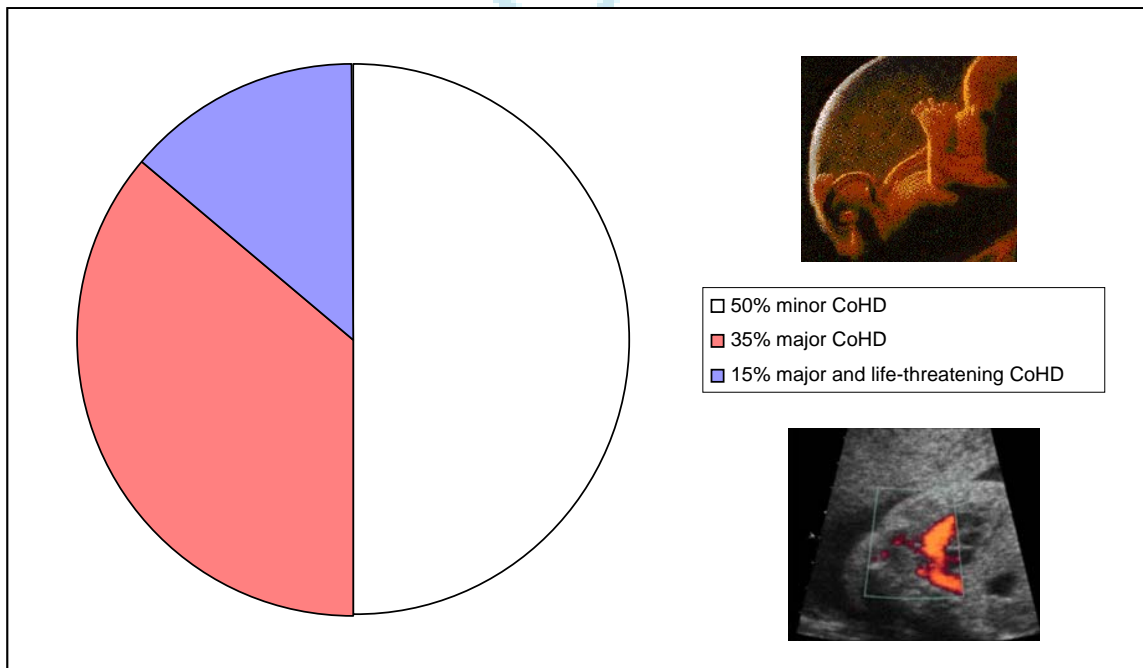


Figure 1. How “heart babies” are affected by CoHD (percentages)

Prospects for babies with Congenital Heart Disease are good

The majority of babies have isolated CoHD, with no genetic associations and their outcome is good. Surgical improvements mean that 80-85% of children born with CoHD now survive to adulthood (BCS Report, Heart, 2002) and in 2003 there were at least 150,000 adults living with CoHD in the UK (BHF report, 2003).

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Prenatal detection is important

Studies show that if CoHD is detected before birth, there are significant benefits for babies, their families and for medical services around the time of birth and in the first year of life (1).

Prenatal diagnosis and appropriate treatment may prevent the devastating consequences of early circulatory collapse, such as brain damage.

Prenatal detection – a postcode lottery?

Routine ultrasound screening has been offered to pregnant women for over twenty years. In 1995, prenatal detection of CoHD for the UK was estimated at 23% (2).

The latest available data derived from children with major CoHD indicates that approximately 30% receive a prenatal diagnosis across the UK (3).

This figure of 30% should be viewed with caution – being derived after birth and with caveats about data availability on the CCAD website. Nevertheless, we welcome this initiative in bringing prenatal detection and diagnosis into the spotlight.

There continues to be wide variation in detection rates throughout the UK, although only average data for each Strategic Health Authority is available at present and there is likely to be wide variation within each Strategic Health Authority region.

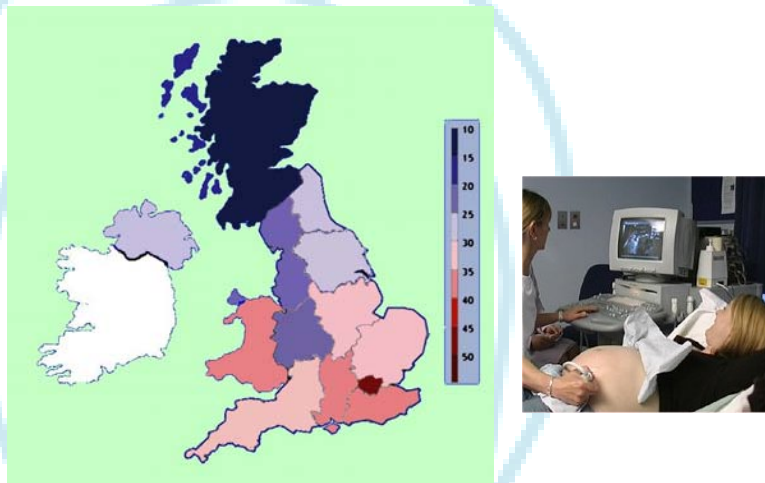


Figure 2. CCAD antenatal diagnosis derived from major CoHD in childhood by Strategic Health Authority, latest available data (3)

REFERENCES

- (1) “Delayed diagnosis of congenital heart disease worsens pre-operative condition and outcome of surgery in neonates”, Brown K et al., *Heart*, 2006;92:1298-1302
- (2) “Current and potential impact of fetal diagnosis on prevalence and spectrum of serious congenital heart disease at term in the UK”, Bull C, *Lancet*, 1999 Oct 9;354(9186):1242-7
- (3) Central Cardiac Audit database (CCAD), www.ccad.org.uk/002/congenital.nsf/vwContent/Antenatal%20Diagnosis (accessed Feb. '10)

EXECUTIVE SUMMARY

1. Integrated Care Pathway

Prenatal diagnosis and appropriate treatment will improve outcomes for most babies with congenital heart disease (CoHD). It may also prevent the devastating consequences of early circulatory collapse, such as brain damage.

To achieve this, it is vital that there is a national approach to improve the detection, referral, diagnosis, management and audit of “heart babies” in pregnancy.

We recommend an integrated prenatal cardiac care pathway with recognised fetal cardiology expertise, similar to that published by the Department of Health, together with the Grown Up Congenital Heart Patients Association, GUCHPA, for Specialist GUCH Consultants.

This should be supported by an integrated approach to congenital cardiac audit, including prenatal data. This should be nationally funded and independent of political change.

2. Improving detection, referral and audit

The detection and subsequent referral of CoHD involves sonographers and midwives, in obstetric ultrasound departments, supported by obstetricians and radiologists.

We recommend an integrated approach to training, accreditation and ongoing education and minimum national standards of ultrasound screening for CoHD at the fetal anomaly scan.

We recommend that every maternity hospital has a health professional with expertise in fetal cardiology, who can provide input in local decision making about screening and referral.

We further recommend investigation into ways to improve overall screening, referral and audit pathways (e.g. through specialist training, telemedicine, feedback from cardiac centres).

3. Adopting standards: The 5 Transverse Views

We recommend a systematic way of screening the whole fetal heart that is proven, practical and comprehensive.

The 5 Transverse View protocol provides an overview of the fetal heart and circulation with a minimum of sweeps and changes in orientation. With suitable training and support, it has been shown to improve detection of CoHD at screening, without adding significantly to the time required. For details refer to: <http://www.tinytickers.org/sonog5vprotocol.html>.

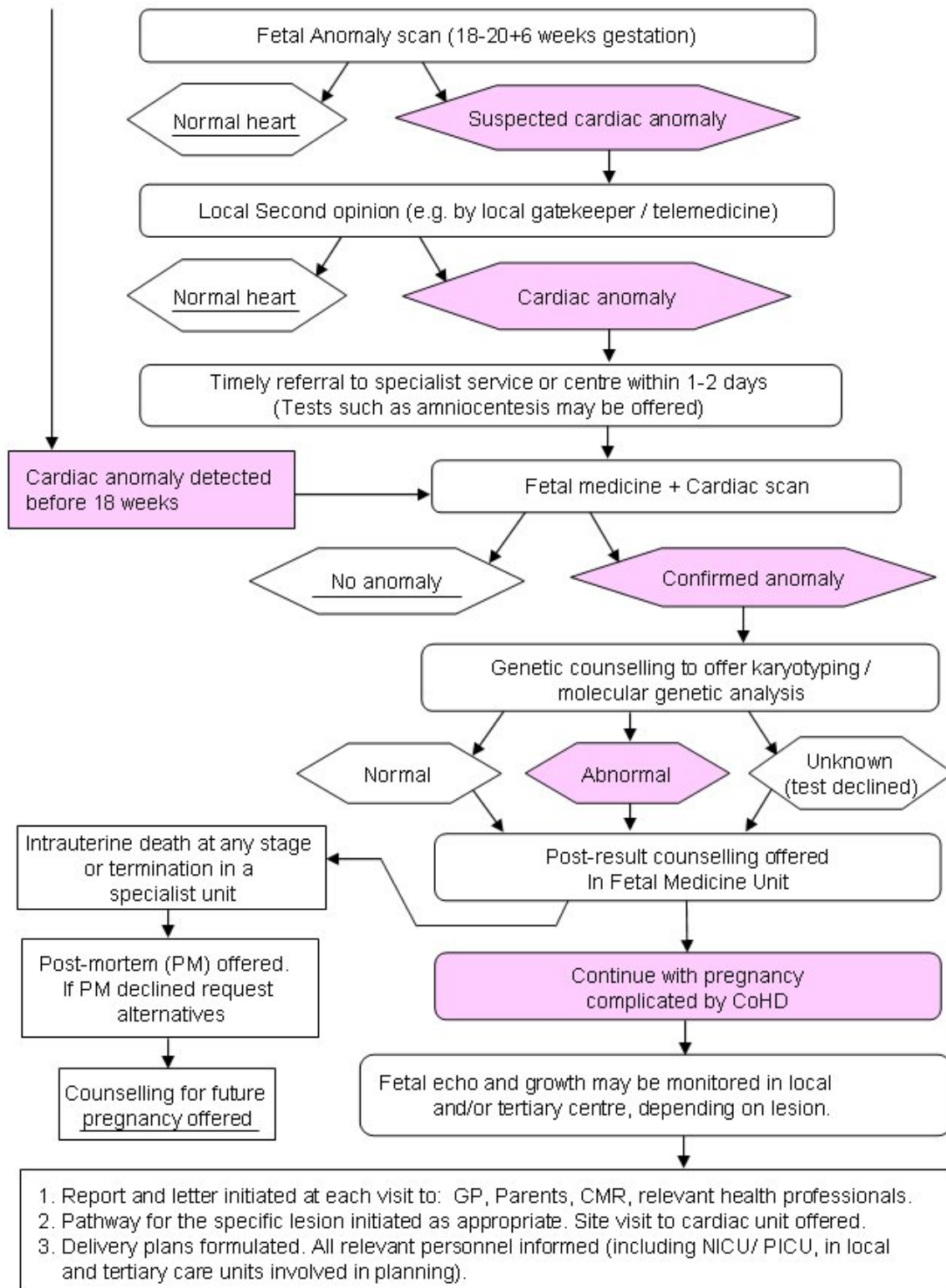
4. Specialist training: expertise in fetal cardiology

Prenatal CoHD should be recognised as requiring expertise that differs from postnatal (paediatric) cardiology, general fetal medicine or obstetrics. In this document we refer to health professionals with this expertise as fetal cardiology specialists.

These specialists should be an integral part of the care pathway, with recognised training and accreditation – for example, see the guidelines published by the European Association of Paediatric Cardiologists (AEPC) and International Society of Ultrasound in Obstetrics and Gynaecology (ISUOG).

Reference our full document: www.tinytickers.org/infoprof_links.html.

SUMMARY OF RECOMMENDATIONS



Map 1: Recommended care pathway for prenatal congenital heart disease

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FETAL ANOMALY SCREENING

The fetal anomaly scan is offered to most pregnant women at 18 to 20⁺⁶ weeks' gestation. This includes an examination of the fetal heart for signs of congenital heart disease (CoHD).

In the UK it is not common to screen for heart disease before 18 weeks. However, some centres do offer this and if a major cardiac anomaly is detected and a woman continues with her pregnancy, she will usually be offered specialist care – either in a Fetal Medicine Unit, or by specialists who provide a fetal medicine service locally.

In Map 1, we show a “Recommended care pathway for prenatal congenital heart disease” and this begins with detection at the fetal anomaly scan or before.

1) Standardised cardiac training for routine fetal anomaly screening

Less than 30% of babies born in the UK with serious heart malformations have a prenatal diagnosis (serious CoHD is defined as requiring surgery or intervention in the first year of life).

Early detection has many benefits for babies, their families and for medical services because it allows appropriate prenatal monitoring, a planned delivery, and avoids emergency situations where babies with CoHD can become ill.

We recommend standardised training and practical assessments in the workplace to improve detection rates for CoHD at the fetal anomaly scan.

We also recommend screening the whole heart - see the 5 Transverse Views, in the Executive Summary.

2) Sufficient time for screening

Many screening departments do not allow sufficient time for scanning, associated paper work and audit, which impacts on the quality of screening.

We recommend that the standard appointment time for the fetal anomaly scan should be no less than thirty minutes. If trainee staff are present this time should be increased further.

3) Adequate electronic systems and audit

Departments that do not have electronic systems usually have less effective audit.

We support the use of electronic audit and ideally digital clip storage of all screened hearts, to improve quality of audit, training and feedback, but this is still not practised in many units.

4) Conducive environment

A calm environment in which to perform a complete examination will aid communication between the health professional and the pregnant woman.

It should be explained to pregnant women that the fetal anomaly scan is a medical screening examination and not a social viewing of the baby by family and friends.

We recommend that where possible child care arrangements should be implemented.

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REFERRAL

5) Reducing automatic referral of “high-risk cases”

“High risk cases” are often automatically referred for a cardiac consultation. These include maternal diabetes, certain drugs, “identical” twins, a family history of CoHD and increased Nuchal translucency.

Numbers of specialists are likely to remain the same, and as prenatal detection increases, demands on specialists will also increase, so automatic referral is not helpful.

We acknowledge that the risk of CoHD in these groups is slightly higher, c. 3%, than the general population. However, with appropriate improvements in screening, we recommend that all pregnant women be scanned for congenital heart disease within the existing fetal anomaly scan. If CoHD is not found, an automatic referral is not necessary.

6) Timely referral

When cardiac views cannot be obtained women are often reviewed up to 2 weeks later.

When cardiac views cannot be obtained we recommend review within 48 hours by an experienced colleague and referral to an appropriately trained specialist if a “normal” heart cannot be seen, as this often indicates an anomaly.

7) Referral via Telemedicine

Access to specialists for triage of cases or second opinion can be facilitated by a Telemedicine link and this can reduce delays and transport costs.

We recommend investment in Telemedicine – especially for remote areas.

Appropriate on-going training of local sonographers and obstetricians, and dedicated specialist resources is essential.

Cost savings should be recognised and used to maintain the service.

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DIAGNOSIS, COMMUNICATIONS AND AUDIT

8) Specialist diagnosis and communications with parents

Detailed diagnosis of CoHD is required for accurate counselling.

Detailed diagnosis of CoHD and subsequent counselling should come from a specialist working in cardiology (i.e. a fetal cardiology specialist, see 9) and supported by an experienced, multi-disciplinary team.

Parents should be made aware of the value of knowing the chromosomes (karyotype) to assist in the planning of mode of delivery, monitoring in labour and decision making in the case of fetal distress as well as postnatal management.

9) Effective communication between health professionals

Congenital heart disease is a complex disease requiring a team approach.

Effective communication between team members, who often operate across multiple sites, is vital to deliver a high standard of care.

Prenatal CoHD should be recognised as requiring expertise that differs from postnatal (paediatric) cardiology, general fetal medicine or obstetrics. Such “fetal cardiology specialists” should be an integral part of the care pathway, with recognised training and accreditation (see guidelines from the European Association of Paediatric Cardiologists, AEPC, and International Society of Ultrasound in Obstetrics and Gynaecology, ISUOG).

Cardiac liaison nurses have an important role in collecting information, discussing this with parents, and providing feedback to the team.

10) Investment in audit and good IT support

Without good audit it is virtually impossible to manage an integrated care pathway. Initiatives such as the CCAD database show how audit can improve the quality of service.

Good quality audit, supported by appropriate IT systems, is vital to improve the integrated care pathway and should include prenatal records (see 11).

11) Access to prenatal records

Currently copies of prenatal letters, reports and delivery and management plans are sent to a variety of locations: mother’s hand-held notes, a mother’s file in the local maternity hospital, the tertiary neonatal unit and paediatric cardiac intensive care unit at the cardiac centre.

This paper trail does not appear to be an effective means of communication.

We recommend a record of all prenatal correspondence and ultrasound scan reports, be held online (e.g. on a secure web-based system, supported by suitable IT resources), for access by the major cardiac centres and maternity units where the baby may be treated in the case of a planned or unexpected delivery.

ON-GOING MONITORING AND MANAGEMENT

12) Ongoing monitoring and management

The benefits of prenatal detection of CoHD include: better care for mothers, appropriate monitoring during pregnancy and planning for a safer delivery in the right place at the right time.

Although it is not always possible, we recommend a woman has a fetal cardiology and fetal medicine obstetric assessment in the same place, as this minimises visits to hospital and improves communication between specialists, including referring obstetricians and sonographers.

At each consultation, sufficient time should be given to discuss management plans and new findings with the family.

13) Preparation for delivery

The cardiac centre is often separate from the maternity hospital.

Parents should be offered a visit to the cardiac unit and neonatal intensive care unit (NICU) before delivery providing the chance to talk to the surgeon, neonatal and cardiac teams, co-ordinated by the cardiac liaison nurse. This will also give the parents a chance to view the parental facilities and plan for their stay in hospital with the new baby if early surgery is required.

14) Specialist post mortem & counselling

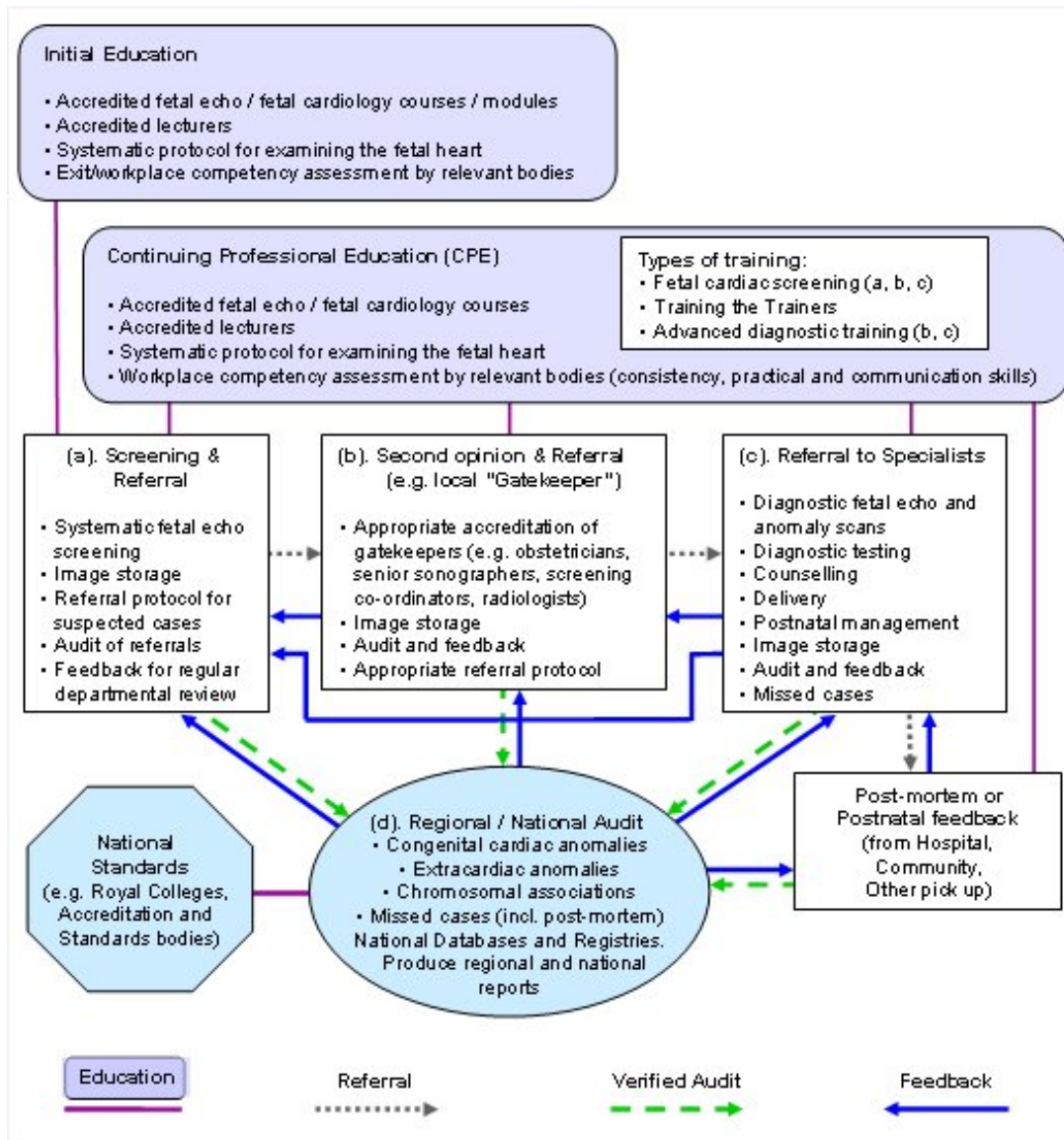
If there is an intrauterine death or termination of pregnancy, a detailed post mortem (PM) of the baby including the heart is vital for counselling before future pregnancies.

We recommend the heart is assessed by an appropriately trained professional with special interest in cardiac pathology (or access to highly specialised professionals if there is no local expertise). If consent for PM is declined, we recommend seeking alternatives, such as consent for medical photographs (for geneticists) and a total body MRI.

Counselling following the end of a pregnancy should be offered to all. The timing is usually about 6 weeks later, once PM results are available and coinciding with a postnatal check by the obstetrician. The purpose is to discuss what happened in this pregnancy, possible reasons and recurrence risks in future pregnancies. An early fetal cardiac scan may be offered for future pregnancies.

PROFESSIONAL PROCESS

This section deals with professional education, audit and feedback, with further recommendations to improve second trimester screening, referral and diagnosis.



Map 2: Recommended professional process for education, audit and feedback

15) Cardiac screening standards: Whilst we support the minimum standards for screening the 4-chambers and Outflow Tracts (NICE Antenatal Screening Update 2008), we recommend a practical, comprehensive and systematic protocol such as the 5 Transverse Views that incorporates the major views of the heart. The protocol includes establishing fetal lie, sweeping up the fetal chest to identify structures and trace cardiac connections

More details: <http://www.tinytickers.org/sonog5vprotocol.html>

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The inclusion of the important 3 Vessel and Tracheal View (View 5) allows detection of important, usually life-threatening lesions involving the aortic and ductal arches (duct-dependent lesions) which comprise 15% of major CoHD (Fig 1).

16) Workplace assessment: To improve CoHD detection rates and maintain that improvement, we recommend practical workplace assessment and continued professional education for all health professionals involved in fetal cardiac scanning, including training, screening, referral, diagnosis and management.

17) Improving audit: Local audit is a vital part of improvement and there should be adequate investment in coding and storing data, including clips, which can then be reviewed to further improve screening or identify training needs.

18) Local second opinion: We support local, expert second opinion - available on the same day as screening - whereby individuals, with specialist training, examine suspected cases before referral to improve education and quality locally.

19) Creating ownership: We recommend focus on developing good leadership and a sense of responsibility and ownership, supported by a learning culture and by good communication, audit and feedback.

20) Increasing resources: Skills for Health proposed that other health personnel be re-trained in obstetric ultrasound screening to address recruitment and resource issues.

Having more resources has other benefits - for example, sonographer sessions can be varied to improve performance and morale.

In a sub-speciality such as fetal cardiology where there are few specialists, limited resources must be used efficiently. One model is the development of fetal medicine obstetricians specially trained in fetal cardiology that triage referrals from local obstetric hospitals and are supported by the congenital heart disease unit/service.

NATIONAL CONGENITAL CARDIAC AUDIT

A national audit of prenatal and postnatal congenital cardiac data will inform the development of the care pathway, enable assessment of screening, identify needs and highlight systemic problems (e.g. low detection rates, delays in referral).

21) Improving patchy CMR coverage:

Congenital Malformation Registers (CMRs), seem to be the best source of prenatal congenital cardiac data, but their existence is reliant on local funding, which is unreliable. A recent example is the demise of NW Thames CMR, which covered c. 47,000 births (or about 8% of UK births).

CMRs gather local pregnancy data on fetal malformations and submit it to national bodies such as Office of National Statistics (ONS) and EUROCAT. They are aggregated via **BINOCAR** (British Isles Network of Congenital Anomaly Registers, www.binocar.org).

National coverage by CMRs is currently about 45%. Ascertainment of prenatal data is poor where there is no CMR.

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To address these issues, we recommend a suitable approach to funding to achieve national CMR coverage and provide a high quality, accurate comprehensive audit.

22) Standardised data collection is desirable to permit amalgamation of data across different registers (e.g. using definitions published by the coding committee) and bridging prenatal and postnatal data. A specialist group should consider the likely areas within the dataset where this may be problematic and formulate recommendations that will be useful for anyone setting up or modifying an anomaly register.

23) Collection of congenital cardiac data should begin before birth, so integration between BINOCAR (local CMRs) and Central Cardiac Audit Database (CCAD) is essential.

We recommend an investigation of how local CMR data and national (cardiac centre and CCAD) data can be made available to improve the service.

24) Enabling National Tracking: Recording prenatal diagnoses (confirmed by fetal cardiology specialists) and postnatal maternal demographics can assist in tracking missed cases and can provide feedback to prenatal screening services in local hospitals, to improve detection rates.

It is essential to make better use of the National Tracking Service which allows one to track via an NHS number. If the cost for this, from one government department to another, creates an artificial barrier to greater use, this should be resolved through a national, integrated approach.

