Br Heart J **(Supplement)** 1994;71:20–27

Screening for fetal cardiac malformations

Jonathan Wyllie, Christopher Wren, Stewart Hunter

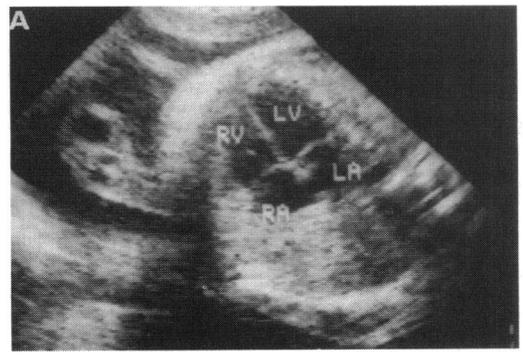
Congenital heart disease (prevalence 8 per 1000 live births) is the most common congenital abnormality.[1 2] It accounts for almost half of perinatal and infant deaths caused by congenital malformation.[3–5] For some time it has been possible to diagnose severe congenital heart disease in the fetus at 18–20 weeks' gestation. This gives the options of counselling, planned delivery, intervention, or termination. The technique is well established but the expertise is available in fewer than 10 centres in the United Kingdom. Anomaly screening by ultrasound of all pregnancies at 18–20 weeks' gestation, including a cardiac assessment, has been advocated[6] and is being implemented. This proposal is not universally accepted.[7] Implementation of such a policy needs careful monitoring and auditing from the start; this did not happen with most previous screening programmes, many of which are only now starting to be evaluated critically. We examine the implications of fetal cardiac screening of all pregnancies, the evaluation of such screening, and our experience of both in the Northern Region.

Fetal echocardiography

Fetal echocardiography has undergone major development in the past 20 years, largely as a result of improvements in ultrasound imaging. The first description of fetal echocardiography was based on the M mode technique.[8] After the development of cross sectional imaging the appearances of the normal heart at 16–18 weeks' gestation were described in the early 1980s.[9 10] In trained hands, fetal echocardiography can now demonstrate most congenital heart lesions at 18–20 weeks.[11–15] Notable exceptions among common defects are the atrial septal defect, because of the extreme thinness of the atrial septum in the fetus, and the arterial duct, which is a normal finding.

In the fetus the heart remains a difficult structure to image in detail. Even with high quality ultrasonography the approach remains indirect and is compromised in adverse situations such as twin pregnancies, oligohydramnios, and maternal obesity. Polyhydramnios increases fetal mobility whereas at advanced gestation or in the presence of oligohydramnios the fetus may be fixed in an unfavourable lie.

Assessment is mainly based upon the tomographic sections used in paediatric echocardiography: the four chamber, left ventricular long axis, ventricular short axis, aortic arch, and short axis pulmonary artery-duct views. Cross sectional imaging is now the main imaging mode but M mode is still used for the analysis of arrhythmias and ventricular wall and septal thickness and assessment of valvar function.[16–18] The single most valuable view of the heart is the four chamber view centred on the atrioventricular junction (fig 1). It is the best way to assess the relative sizes of the chambers, the anatomy of the atrioventricular valves, and the presence of pericardial fluid. It is obtainable in more than 95% of pregnancies[15 19] by imaging a transverse section across the fetal thorax above the level of the diaphragm. The right ventricle is blunt ended and anterior with a more obvious moderator band than in postnatal life. The following features must be assessed: (*a*) the heart should occupy not more than a third of the fetal thorax; (*b*) there should be two atria of equal size; (*c*) there should be ventricles of equal size that contract equally briskly; (*d*) the two atrioventricular valves should meet the atrial and ventricular septum at the crux, giving the appearance of an offset cross; (*d*) the foramen ovale should be present; (*f*) and the ventricular septum should appear to be intact.[13] More experienced operators will be able to identify connections of the pulmonary veins joining the left atrium (fig 2).



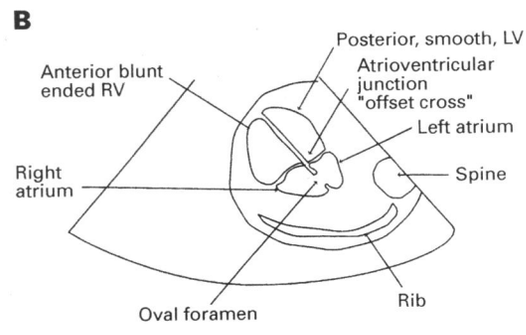


Figure 1 Four chamber view of the fetal heart. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Department of Paediatric Cardiology, Freeman Hospital, Newcastle upon Tyne
J P Wyllie
C Wren
S Hunter

Correspondence to:
Dr S Hunter, Department of Paediatric Cardiology, Freeman Hospital, High Heaton, Newcastle upon Tyne, NE7 7DN.

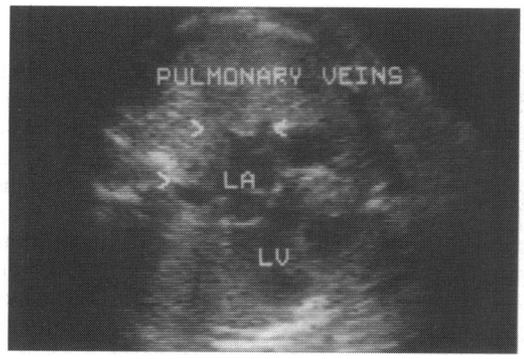


Figure 2 Fetal pulmonary veins entering (arrow heads) the left atrium. See legend to fig. 1 for abbreviations.

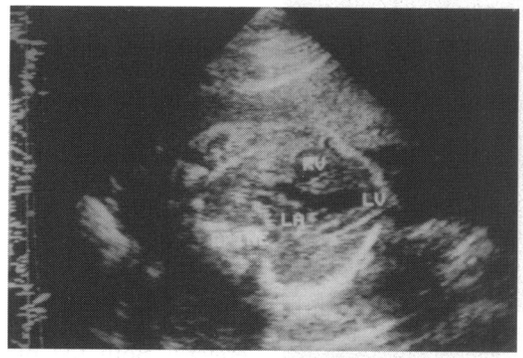


Figure 3 Long axis view of fetal heart.

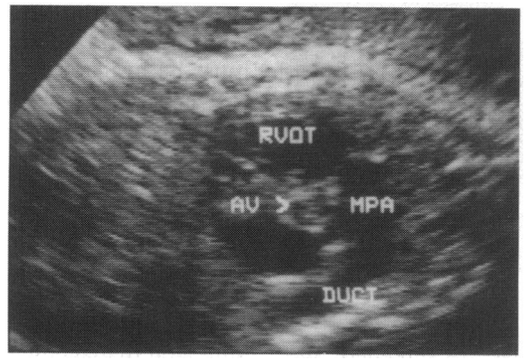


Figure 4 Short axis view of fetal heart. AV, aortic valve; MPA, main pulmonary artery; RVOT, right ventricular outflow tract.

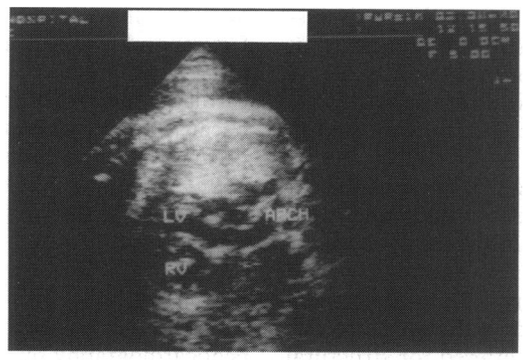


Figure 5 Fetal aortic arch connecting to the left ventricle.

It is more difficult to evaluate the outflow tracts. Continuity between left atrium, left ventricle, left ventricular outflow tract, and aorta can be demonstrated by obtaining a traditional long axis view (fig 3). The normal relation between the aorta and pulmonary artery is best seen in the short axis plane (fig 4). Lastly when the aortic arch is visualised care must be taken not to mistake it for the ductal arch, which is also easily imaged. The aorta is distinguished from the duct by identifying its characteristic arch, the presence of head and neck vessels, and tracing its origin to the left ventricle (fig 5).

Screening for heart disease in the fetus

Obstetric ultrasound scanning is performed in more than 80% of pregnancies in the United Kingdom.[20] The timing varies but in the Northern Region an "anomaly scan" is done at 18–20 weeks' gestation in 91% of pregnancies (results of a questionnaire). Several groups have shown this is the optimum time for cardiac assessment from the transabdominal approach.[9–14] The most skilled fetal cardiac ultrasonographers are paediatric cardiologists and specialist fetal echocardiography technicians. It would be a counsel of perfection that such an expert should perform a screening scan in all pregnancies. Fermont *et al*[21] estimated that a specialist fetal echocardiographer would have to perform 300 hours of routine obstetric scanning to detect one serious cardiac defect (based upon an incidence of 4 per 1000 pregnancies and two scans per pregnancy). Therefore, to cover the United Kingdom, where it is usual to perform only one anomaly scan, one expert working full time, would be needed for every 1800 pregnancies—that is, about 400 fetal cardiac specialists. The lack of skilled personnel and the enormous cost rule out this approach.

The obvious alternative approach is to incorporate the four chamber view into the routine obstetric assessment and provide extra training for obstetric sonographers.[19 21–24] This should identify most of the complex defects and detect malformations in up to 2 per 1000 pregnancies.[22–26] Because anomaly scanning is already so widely performed there is an opportunity to screen all pregnancies for major forms of congenital heart disease,[13] if the four chamber view becomes part of the routine anomaly scan. When a normal four chamber view cannot be obtained or an abnormality of the chambers is recognised in this view (fig 6) the case should be promptly referred to a specialist centre. In addition high risk pregnancies should be referred directly for an expert assessment. At present we routinely scan fetuses because of (*a*) a family history of congenital heart disease, (*b*) maternal diabetes mellitus, (*c*) identification of a non-cardiac abnormality, (*d*) non-immunological hydrops, (*e*) exposure to teratogens, and (*f*) fetal arrhythmias. As yet, however, not all such high risk pregnancies in the Northern Region are referred for assessment. Even if they were they would account for only 5–6% of cases of congenital heart disease[26 27] and most defects would be detected from routine scans.[22 23 26–29] Because most obstetric units might expect to see fewer than 10 cardiac malformations per year, feedback and continued training are vital.

The four chamber view will detect most, though not all, serious heart malformations, (table 1). In addition it may show changes in chamber size that are not associated with structural heart disease. The recognition of any abnormality necessitates a rapid referral for a definitive paediatric cardiological assessment. Antenatal diagnosis of a cardiac anomaly obviously causes parental anxiety and highlights the need for informed decisions by

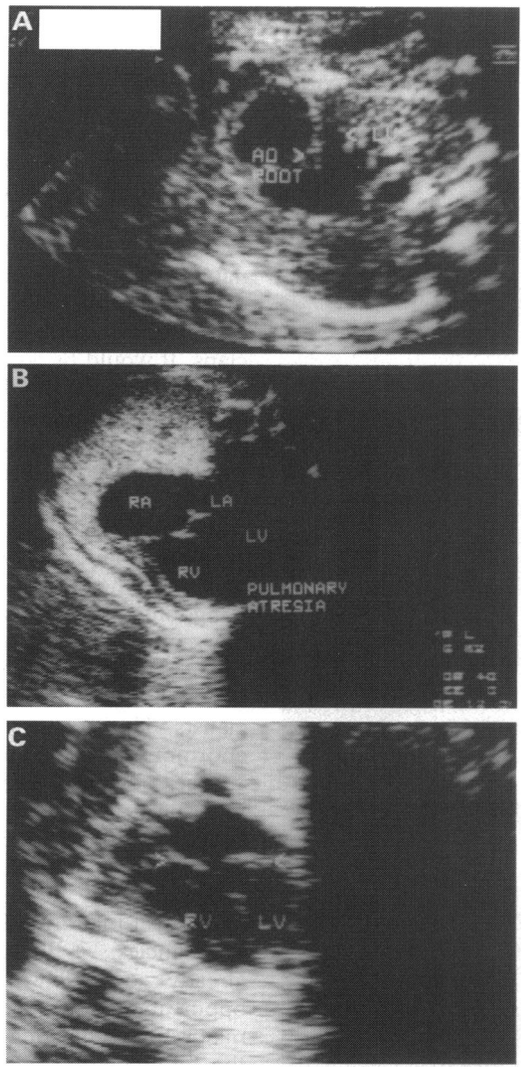


Figure 6 Four chamber views showing (A) hypoplastic left ventricle (Ao, aortic root), (B) pulmonary atresia with large right ventricle, and (C) complete atrioventricular septal defect with absence of offset cross.

parents before pregnancies are screened. We are aware that many mothers still regard their ultrasound examination as purely an assessment of gestation. Mothers still present to us unaware of the reason for their referral. More effort must be made to keep parents fully informed of their choices before a pregnancy is screened.[30 31]

Once a diagnosis is made there must be the infrastructure for counselling and supporting parents, who must be told by the obstetrician or midwife about the implications of an abnormality scan before it is performed. Termination of pregnancy is the only effective intervention that can be offered for structural congenital heart disease. Antenatal diagnosis of congenital heart disease offers, in addition, the possibility of genetic counselling, therapeutic intervention, and choice of location and mode of delivery. We have seen no evidence that delivery of babies with known isolated congenital heart disease in a tertiary referral unit improves outcome and such delivery is not our policy. However, after delivery appropriate medication may be started (for example, prostaglandin) before transfer and babies should reach the centre in optimal condition.

Only a few therapeutic interventions are possible in utero at present[31] and the as yet uncertain natural history of cardiac anomalies in fetal life makes appraisal of such interventions difficult. Fetal arrhythmias are an exception and they can be successfully treated by maternal medication.[33 34] Termination of pregnancy remains the major intervention resulting from a diagnosis of congenital heart disease in the fetus before 23 weeks' gestation. This is what up to 75% of parents chose.[22 26 36–38] This will certainly reduce the prevalence of serious cardiac malformations if implemented nationally but termination at 20–23 weeks is often more distressing than at an earlier gestation. Resources for counselling after termination are inadequate and many parents feel unsupported.[39] In future transvaginal echocardiography may offer an earlier diagnosis[40 41] but it may never be a practical routine alternative despite the advantage of termination at 12–14 weeks.

Table 1 Lesions potentially detectable by four chamber scan

- Hypoplastic left heart
- Pulmonary atresia with intact septum
- Double inlet left ventricle
- Mitral atresia
- Tricuspid atresia
- Critical aortic stenosis
- Critical pulmonary stenosis
- Atrioventricular septal defect
- Ebstein's anomaly

General philosophy of screening

The value of any screening test is assessed in terms of simplicity, acceptability, accuracy, repeatability, specificity, sensitivity, cost, and effect.[41] Both the pilot trials and the established screening test should be assessed according to the World Health Organisation guidelines and adequate methods to do this must be available.[43] Many screening policies that have long been thought to be invaluable are now being reassessed and abandoned.

SIMPLICITY

The four chamber view is the easiest of the fetal cardiac views to obtain and with appropriate training it can be incorporated into a routine obstetric abnormality scan at 18 weeks, without requiring major changes to antenatal management.

ACCEPTABILITY

Ultrasound is widely accepted as safe for both mother and fetus,[44 45] and screening for fetal cardiac malformation is widely accepted because of the reassurance it offers in most pregnancies.[46] Parents should be told more about the choices presented when a malformation is diagnosed at 16–20 weeks' gestation.

ACCURACY

Many studies have shown the accuracy of fetal echocardiography by skilled operators.[9–14] The four chamber view is a reliable method of identifying an absent valve or chamber or a lesion causing asymmetry.[18 19 22 23] These are, by definition, the more severe and "uncorrectable" congenital heart anomalies.

REPEATABILITY

Both four chamber screening and detailed echocardiography gave consistent results in limited trials.[14 15 19 21 22] A four chamber view is obtainable in 90–98% of pregnancies[15 19 22] but few studies have assessed the usefulness of this view when it is obtained by non-specialists. Fermont *et al*[21 25] and Allen *et al*[13 22 36] found it consistent in detecting serious heart

lesions in repeated trials. They did, however, require close cooperation between ultrasonographers, obstetricians, and fetal cardiologists.[21 22 25]

SPECIFICITY

Specificity is the likelihood that a screening test will be negative when the fetus does not have an abnormality—that is, the ability of the test to recognise a normal heart. It is expressed as the number of normal individuals whose scan is negative in relation to the number of normal individuals in the population being screened. The specificity of fetal cardiac screening is likely to be high because large numbers are screened and few have congenital heart disease. Even if 90% of those referred for detailed evaluation proved to be normal the specificity would still be > 98%. The reported results of several trials accord with this figure.[12–15 19 21 22]

Sensitivity, costs, and effect are more difficult to assess.

SENSITIVITY

Sensitivity is the likelihood that the test will be positive when the individual scanned has the abnormality—that is, the ability of the test to recognise a cardiac malformation. It is expressed as the number of congenital heart abnormalities detected in relation to the number of abnormalities in the population being screened.

It is not difficult to assess the sensitivity of screening in tertiary centres. It is uniformly high[14 15 19 21–23] in this selected population. None of the studies of screening by specialised echocardiography is based upon true population data. All rely on assumptions about referral patterns[14 23] or are based on single hospitals.[15 19] Even the populations in the two best studies of four chamber screening are poorly defined, with a sensitivity of 77% and 92%.[19 22] It is much easier to follow up all the fetuses with positive scans than to identify all cases of congenital heart disease presenting postnatally in the same population. For a true assessment of sensitivity it is essential to know this denominator. Typically the population boundaries and referral patterns are unclear and it is difficult to connect infants and mothers in different databases when they have different surnames. Furthermore, up to a third of infant deaths from very severe congenital heart disease occur before diagnosis, which is made only after death.[47] These children are part of the denominator that is used to calculate sensitivity and data on them are available only if formally collected. The Northern Regional Fetal Abnormality Survey collates such data[3] making the region ideal for studies of the sensitivity of any screening test for fetal abnormality.

COSTS

It is difficult to measure accurately the cost of screening. The addition of four chamber screening to the obstetric abnormality scan does not require capital outlay on equipment. Most obstetric units have adequate machines, and all pregnant women have access to such facilities in the Northern Region. Extra staff will be needed, but many fewer ultrasonographers than forecast by Fermont *et al*[21]—about 35–55 for the United Kingdom (rather than 400), assuming that a four chamber view takes 2–3 minutes (30 hours per week scanning for 45 weeks for 750 000 pregnancies).

Obstetric ultrasonographers are already in place but they will require extra training. We found that, although initial training had an effect, continued input and feedback to maternity units was needed. The best results in the United Kingdom were obtained in a professorial department of fetal cardiology largely supported by charitable funding.[22] This is not practical for most regions, but to have any impact there must be feedback to and continued training and support for ultrasonographers at individual peripheral units. In other words audit must start when the screening scheme is implemented. Ultrasonographers who see two or three lesions a year find it less easy to remain enthusiastic than those who see two or three lesions a month.

To implement a regional screening programme we recently appointed a full-time regional specialist technician in fetal echocardiography. We will review progress in 1996. This approach is likely to cost less than funding further professorial units of fetal cardiology.

Costs are also incurred in staffing tertiary referral centres. At present, fetal cardiology training is not an essential part of training paediatric cardiologists in the United Kingdom: it will be in the future.[48] Current resources could not sustain a truly national screening programme.

It is difficult to estimate the saving made as a consequence of termination of pregnancies or the arrival at the specialist centre of infants with severe congenital heart disease, such as hypoplastic left heart syndrome, in better condition. With termination the costs of care and surgery are avoided whereas improving the condition of infants with congenital disease could increase costs. A cost benefit analysis based upon previous work is needed before national screening is implemented. The emotional costs can only be judged by parents: they are difficult to assess formally.[39] We found that most parents who have already had an affected child opt for antenatal diagnosis. They base their choice on their experience but with no knowledge of the reality of midtrimester termination.

EFFECT

An assessment of the effect is a prerequisite of a cost benefit analysis. Several studies have predicted far reaching effects on the future practice of paediatric cardiology.[15 22 26] These were based on the perceptions of prevalence of local or single centres or large North American studies of prevalence.[15] The weakness of such an approach is that the number of minor abnormalities is underestimated as is the number of major abnormalities that escape detection before death. Prospective studies have

relied upon symptomatic presentation rather than postnatal examination of all children.

A contemporaneous, population based register of congenital heart disease would be needed to evaluate the effect accurately. In its absence there is evidence for the potential to detect a large proportion of severe congenital heart disease at 18–23 weeks' gestation. Only termination has a proven effect. Sharland *et al* [22] found that 75% of parents opted for termination: others quote a lower figure.[26 38] This would reduce subsequent perinatal and infant mortality. Studies are in progress in the North East and in London to assess this effect.

Fetal cardiac screening in the Northern Region

The aims of this study were:

- To evaluate antenatal cardiac screening in maternity units and the regional referral centre.
- To assess the effect of training obstetric ultrasonographers in obtaining a four chamber view at 18–20 weeks.

The Northern Region is demographically well defined with a population of 3 076 600 and about 40 000 births a year. All cases of congenital heart disease from 15 of the 16 health districts are seen at the Freeman Hospital or peripheral clinics attended by one of three paediatric cardiologists. South Cumbria is the only district that refers congenital heart disease out of the region and we excluded it from the study, though some fetal abnormalities are referred to Newcastle. Apart from this district there is no extra-regional referral of cases of infant congenital heart disease.

The Regional Fetal Abnormality Survey[3] and the Perinatal Mortality Survey[49] record information about all congenital malformations, terminated pregnancies, and stillbirths as well as infants dying before diagnosis of congenital heart disease. They also provide information on the regional birth rate, numbers of stillbirths after 20 weeks' gestation, and the number and outcome of therapeutic terminations. Their methods are well validated.[3 49]

In the Northern Region there are 18 maternity units plus four run by general practitioners. For some years five of these units gave extra training in obtaining the four chamber view (SH). These five units account for just under half of the region's deliveries.

A questionnaire was sent to each maternity unit to gather information about the ultrasound equipment in use and the proportion of fetuses scanned at 18–20 weeks' gestation. This enabled us to estimate the actual number of fetuses scanned at this gestation in all maternity units in the region between August 1990 and July 1992.

Information on the reason for referral and the outcome of all fetal scans performed at Freeman Hospital between August 1990 and July 1992 was obtained prospectively. This period was chosen as representing the two years during which scans were performed at 20 weeks' gestation on fetuses due to deliver in 1991–92 inclusively (fig 7).

We identified all children with congenital heart disease born alive between January 1991 and December 1992 who presented in infancy to the Freeman Hospital or to peripheral clinics. Case ascertainment continued until the end of 1993 to ensure that all infant presentations were included (fig 7). The two collaborative surveys (Northern Regional Fetal Abnormality and Perinatal Mortality Surveys) provided information on children with congenital heart disease that was undiagnosed or unconfirmed before death.[3 49] They were also the source of postmortem data on terminated fetuses, antepartum stillbirths after 20 weeks' gestation, and on children dying of unrelated causes in whom an incidental diagnosis of congenital heart disease was made.

Congenital heart malformations were classified as complex if a valve or a chamber was atretic or hypoplastic, as "significant" when four valves and chambers were present but intervention was required, and as minor if no treatment was required. We excluded preterm babies with patent arterial ducts and babies with cardiomyopathy or postnatal arrhythmias (table 1).

Each cardiac abnormality was further classified as "detectable" or "not detectable" by a four chamber scan performed by an obstetric ultrasonographer at 18–20 weeks' gestation. As already stated, (table 1) a few ventricular septal defects are detectable in the four chamber view. Because the proportion that should be detectable cannot be judged we excluded ventricular septal defects.

RESULTS

The questionaire showed that all pregnant women had access to examination by good quality ultrasound machines and, if booked early enough, were offered abnormality scans. More than 97% of all pregnancies in the Northern Region were scanned (91% between 16 and 20 weeks). In the two years of the study there were 77 262 live births and 77 648 fetuses at 20 weeks' gestation. Therefore, 70 660 pregnancies must have been scanned at 18–20 weeks' gestation (fig 8).

There were 519 referrals for detailed echocardiographic assessment in the regional centre. Figure 9 shows the numbers and reasons for referral. A previous family history of congenital heart disease was the commonest reason for referral but the most productive was an abnormality on a four chamber view found during a routine obstetric scan.

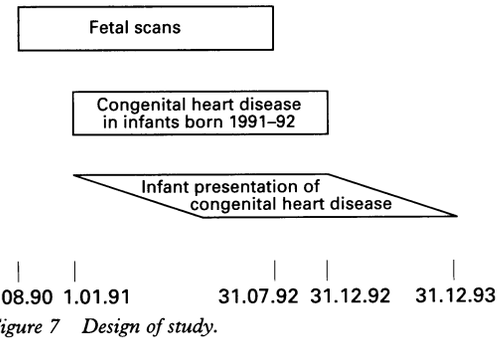


Figure 7 Design of study.

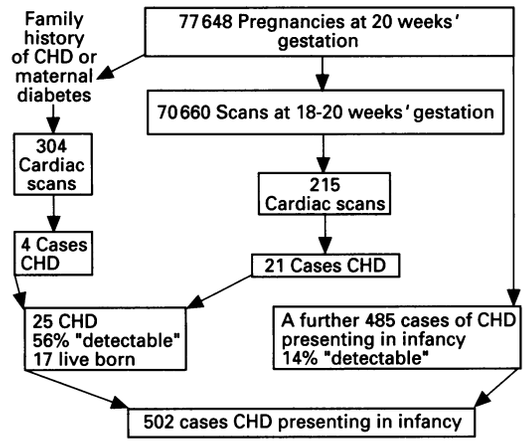


Figure 8 Diagnosis of congenital heart disease (CHD) from 20 weeks' gestation.

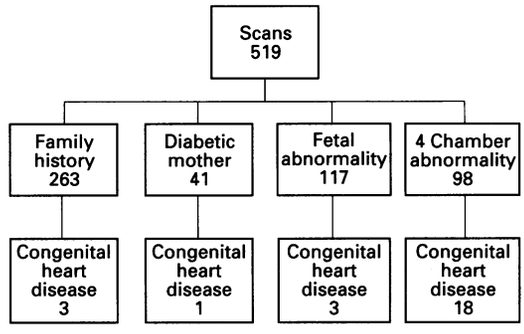


Figure 9 Referral pattern and diagnoses.

Twenty five structural heart lesions (table 2) and 20 non-structural lesions (table 3) were detected in utero during the two years of the study. No false negative diagnoses were made by the regional department. One false positive diagnosis of ventricular septal defect was made and an initial false positive diagnosis of transposition of the great arteries was corrected at a second scan two days later. However, one diagnosis was changed from a large ventricular septal defect to mitral atresia on follow up scan and one other diagnosis was slightly modified by postmortem examination after termination. The initial postmortem examination of one fetus suggested an incorrect echocardiographic diagnosis but a more detailed expert appraisal confirmed the original diagnosis. In two fetuses muscular ventricular septal defects were followed serially and the defects closed as the pregnancy progressed. Postnatally their hearts were structurally normal.

Twelve fetuses had right atrial and right ventricular hypertrophy with tricuspid regurgitation in the absence of any other demonstrable heart abnormality (table 4). Three were associated with congenital diaphragmatic hernias and one of these was found postnatally to have an atrial septal defect. In two of the 12 cases right heart enlargement was confirmed postnatally but subsequently resolved. The poor outcome of isolated pericardial effusions found during this time has already been reported[49] but two cases associated with supraventricular tachycardia were successfully treated in utero.

Of the 25 structural lesions detected 15 were "detectable" by routine four chamber view (table 1 and fig 8) but only 12 of 15 were found before 24 weeks' gestation. After counselling (SH, JPW, and obstetrician) seven (58%) sets of parents elected to terminate the pregnancy (fig 10). The remaining eight malformations were all ventricular septal defects detected after referral from centres of fetal medicine or for assessment because of other abnormalities. Two ventricular septal defects required surgical closure in infancy, two closed antenatally, and the remaining four were small and unlikely to require surgery.

In 1991–92 there were 502 confirmed cases of congenital heart disease in the Northern Region presenting in infancy (6·5 per 1000 live births). Of these malformations 60 were complex, 223 significant, and 219 mild. Eighty two fetuses had "detectable" cardiac malformations in the Northern Region at 20 weeks' gestation but five of these were antepartum stillbirths. Therefore screening detected 18·3% of detectable malformations but only 13% were detected before 24 weeks. There were 60 complex lesions for which no surgical correction was possible. Of these 39 (73%) were potentially detectable: 7 (18%) were found.

Table 3 Non-structural fetal heart abnormalities detected

Abnormality	*No*	*Outcome*
SVT	2	Both treated successfully
Isolated pericardial effusion	6	4 in utero deaths, 1 neonatal death, 1 normal survivor[48]
Right atrial and right ventricular hypertrophy and TR	10	9 normal, 1 intrauterine death
Right and left ventricular hypertrophy	2	1 with ovarian cysts, 1 with renal agenesis
Total	20	–

SVT, supraventricular tachycardia; TR, tricuspid regurgitation.

Table 2 Fetal structural heart disease detected

Lesion	*n*	*Number found < 24 weeks' gestation*	*Terminated pregnancies*
Hypoplastic left heart	5	5	3
Pulmonary atresia, intact ventricular septum	1	1	–
Aortic stenosis	2	–	–
Complex complete atrioventricular septal defect	4	4	4
Double outlet right ventricle	1	1	–
Tricuspid atresia	1	–	–
Ventricular septal defect	9	4	–
Atrial septal defect	2	2	
Total	25	17	7

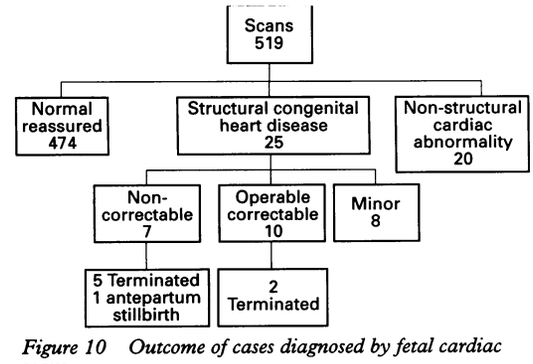


Figure 10 Outcome of cases diagnosed by fetal cardiac scans.

Table 4 Outcome in fetuses with isolated right atrial and right ventricular hypertrophy with tricuspid regurgitation

Outcome	*n*
Normal postnatally	7 (2 CDH)
RAH + RVH with TR resolving postnatally	2
ASD	1 (1 CDH)
Hydrops IUD, structurally normal heart	1
ASD/small VSD	1
Total	12

RAH, right atrial hypertrophy; RVH, right ventricular hypertrophy; TR, tricuspid regurgitation; ASD, atrial septal defect; VSD, ventricular septal defect; CDH, congenital diaphragmatic hernia; IUD, intrauterine death.

Sensitivity

The sensitivity of screening by obstetric ultrasonographers was, as expected, high (> 99%) and this was also true for the regional centre.

Specificity

Specificity was assessed in relation to the numbers of detectable lesions at 20 weeks' gestation. Specificity was 18·3% for obstetric operators and 100% for the regional referral unit. The positive predictive value of an obstetric referral with a suspected abnormal four chamber view was 18·4% but there was considerable variation between units. For some units the predictive value of referral was >70%.

Only two malformations were detected in hospitals where staff had not been trained in obtaining the four chamber view. In these hospitals 43 364 pregnancies reached 20 weeks' gestation (57% of the regional total) and in the five units where additional training had been given 34 284 (43%) reached 20 weeks' gestation. These five units detected 21 anomalies: only 13 were "detectable" by the four chamber view alone. There was great variation even among these five units, with the best detecting 58% of "detectable" abnormalities and the worst none. The most successful were those staffed by people with a specific interest in ultrasonography or those that had a department of fetal medicine. There was no significant difference in the prevalence of "detectable" congenital heart disease between the two types of unit. Of 73 cases, 42% occurred in units where extra tuition had been given and 58% where it had not.

DISCUSSION

Our data suggest that more pregnancies are being screened at 18–20 weeks' gestation in the Northern Region than nationally.[20] The proportion screened is similar to a smaller more recent report from Luton.[51] All units in the Northern Region attempt a four chamber view, indicating that more widespread four chamber screening would need little extra organisation. It would, however, require trained fetal cardiologists, counsellors, and support for parents.

The referral pattern for detailed fetal echocardiography mirrors that found by others.[13 28 29] A family history of congenital heart disease was the most frequent reason but referral with an abnormal four chamber scan was the most productive. It is difficult to assess the value of reassurance given to large numbers of parents who have already had one child affected by congenital heart disease. We have seen increasing numbers of mothers referred because of diabetes mellitus. If all such women in the region were referred for detailed scanning, as has been suggested,[13] there are likely to be an extra 120 detailed examinations annually.

We detected only 18% of "detectable" lesions in the Northern Region over the two years studied. This is much lower than some previous studies but resembles the figures given by most other regions. However, our ascertainment of cases is better than most because of the Regional Fetal Abnormality Survey.[3] This makes our results accurate and population based but makes our detection rate lower than in other regions. However, the existence of the regional survey, the cardiology database, and the fetal scan database permits assessment which begins to fulfil the criteria for the appraisal of a screening test.

The detection rate in referring units ranged from 0% to 58%. Those in which staff had additional training performed much better. It is impossible, however, to draw firm conclusions because these units were also larger and, though there was little difference in equipment, their staff were both more experienced and had a greater interest in fetal medicine. It is likely that these factors were important.

The sensitivity of screening in the Northern Region resembles that elsewhere. It is a function of the prevalence of congenital heart disease. Our specificity, however, was poor. The predictive value of an obstetric referral with an abnormal four chamber view was only 18·4%. Though this is much better than the predictive value of any other referral mode, it is only about half of that quoted generally[19 22 23] and is much less than for other fetal abnormalities.[51] These results come from a whole health region. Others have shown that with the backing of a professorial department[22] or detailed screening in single hospitals[15] the results are good. Some of our units achieved similar detection rates with fewer resources. Our first aim must be to raise the general level of skills to that of the best in the region, concentrating on the four chamber view. We accept that feedback is vital so that ultrasonographers know what they are detecting and what they are missing. To this end, and to improve and continue training, we have appointed a regional perinatal echocardiographer for the next three years. We will report on progress later.

Our results so far would not recommend prenatal cardiac screening to a health economist. Provision of training and an audit system such as we plan in the Northern Region will undoubtedly improve the detection rates throughout the region. More factors other than cost must be considered. The demand for this service is increasing: referrals went up by just under 50% from 1991 to 1992. There must be a full evaluation of the potential for a four chamber screening test, along the lines we have described, to justify future funding. Such a national evaluation has been started under the auspices of the British Paediatric Cardiac Association but data collection is likely to be difficult for reasons already mentioned. As a first stage all centres offering the service must be able to acquire the data we have described. They must also be able to assess the effect on families, which they do not do at present.

Screening for congenital heart disease in the fetus is here to stay because parents want it and because it increases our knowledge about congenital heart disease.[50 52–54] We must determine in what form it should be offered and to what end.

We thank Ms Alison Heads, Northern Regional Perinatal Echocardiography Tutor, for technical help and assistance and all those who contribute to the Northern Regional Fetal Abnormality Survey and Perinatal Morbidity Survey. J P W is supported by a grant from the Sir Jules Thorne Charitable Trust.

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