Fetal Heart Evaluation for all pregnancies: A Proposal

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Congenital heart defects are one of the most common birth defects in neonates and infants.

Prevalence: 6-10/live births (2-3/1000 critical)
The Kerala Scenario

Best human development index in India

Best literacy rates

IMR: 12/1000 live births.

Crude birth rate 14.6

Access to antenatal care 100%

Institutional births 99.5%
CHD as a contributor to infant mortality: Kerala Scenario

• Importance cause of IMR, especially in low IMR areas (4th leading cause of neonatal deaths)

• 60% of critical CHDs die before the first birthday.

• 25% infants deaths in Kerala attributed due to CHD*

* UNICEF - IMA partnership concept note
Severity of CHD: RACHS system

1. ASD
   PDA > 30 d
   CoA > 30 d

2. VSD
   TOF
   TAPVC > 30 d
   BDGS

3. RV-PA conduit
   Fontan
   BT shunt
   ASO
   ALCAPA

4. Complex ASO
   Truncus
   IAA repair
   Double Switch
   Unifocalization

5. Neonatal Ebsteins repair
   Truncus with IAAA

6. Stage 1 Norwood

Simple
Major
Complex
Very Complex
CHD Management: Priorities for limited resource healthcare systems

- More conservative approach to complex CHD with suboptimal outcomes
- Early referral of correctable CHD
- Optimal resource utilization
- Early referral of correctable CHD
Early diagnosis of CHD –
Changing Paradigms

Triaging based on healthcare priorities
Accuracy of fetal echocardiography in the routine detection of congenital heart disease among unselected and low risk populations: a systematic review

Objective To determine among unselected and low risk populations the accuracy with which fetal echocardiography during the second trimester detects congenital heart disease.

Efficacy of Routine Fetal Ultrasound Screening for Congenital Heart Disease in Isère County, France

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Recent literature has revealed different percentages of prenatal detection of congenital heart disease. Therefore we chose to analyse the sensitivity of this screening in Isère county and to scrutinize to which extent factors like severity of the anomaly, extracardiac anomalies and maternal age influence the sensitivity.

This retrospective study covers the period from 1989 to 1995. All the pregnancies with congenital heart disease in Isère county in France were reviewed and analysed, which was possible thanks to our registry of congenital anomalies. In this period, 316 cases were registered in the central database. We obtained an overall sensitivity of 34.8 per cent. By splitting the different malformations into two groups we got a detection rate of 53.7 per cent for major malformations and 26.7 per cent for other abnormalities. This difference is significant. Nevertheless, the sensitivity remains quite low. This result stresses the need for better education of investigators in primary care units, particularly because the prenatal diagnosis of congenital heart disease has a major impact on the outcome of pregnancy, which can be seen in the increased number of abortions in this group. Copyright © 1999 John Wiley & Sons, Ltd.

Prenatal detection of heart defects in a non-selected population of 30 149 fetuses – detection rates and outcome

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Results Of 97 major CHDs, 55 (57%) were detected prenatally, 16% (9/55) prior to, 66% (36/55) at and 18% (10/55) after the routine scan. Forty-four percent (19/43) of the isolated CHDs, 67% (36/54) of those with associated malformations and 48% (11/23) of the isolated ductal-dependent CHDs were detected. Thirty-
Options for management after Prenatal Diagnosis

**Complete Cardiac diagnosis**
- In-utero prognosis
- Extra-cardiac factors
- Critical presentation at birth
- Outcomes Institutional experience
- Costs initial and recurring

**Gestation age**

**Planned delivery and post natal care**
**Planned Comfort Care**
**TOP**

*Based on local, regional and national laws and regulations*
Antenatal screening for CHD:

2 Approaches

Targeted study
“High Risk”

Screen Everyone
Indications for Fetal Echocardiography

10-20% of all CHDs occur in high-risk pregnancies

**FETAL**
- Abnormal 4C view
- Extracardiac anomalies – GIT, spina bifida
- Chromosomal anomalies – VACTERL, Trisomies, Digeorge
- Non-immune hydrops
- IVF/ICSI
- Irregular heart beat
- Increased first trimester NT
- Abnormal cardiac axis

**MATERNAL**
- Maternal CHD
- Maternal auto-immune disease
- Teratogen exposure
- Metabolic disorders – DM, PKU
- Intra-uterine infections

**FAMILIAL**
- Previous child with CHD
- Paternal CHD
- Mendelian syndromes – TS, Noonan’s, Digeorge
Maternal factors

- Diabetes mellitus: Pre-gestational, first trimester GDM, HbA1C > 8.5
- Phenylketonuria (levels > 15 mg/dL)
- Autoimmune disease
- Medication exposure – AEDs, SSRI, NSAID
- Infections
- ART (ICSI/IVF)
Risk of congenital heart defects associated with assisted reproductive technologies: a population-based evaluation

Karim Tararbit\textsuperscript{1}\textasteriskcentered, Lucile Houyel\textsuperscript{2}, Damien Bonnet\textsuperscript{3}, Catherine De Vigan\textsuperscript{1}, Nathalie Lelong\textsuperscript{1}, François Goffinet\textsuperscript{1,4}, and Babak Khoshnood\textsuperscript{1}

Methods and results

We used data from the Paris Registry of Congenital Malformations on 5493 cases of CHD and 3847 malformed controls for which no associations with ART were reported in the literature. Assisted reproductive technologies included inductors of ovulation only, in vitro fertilization, and intracytoplasmic sperm injection. Exposure to ART was higher for cases than controls (4.7 vs. 3.6\%, $P = 0.008$) and was associated with a 40\% increase in the maternal age, socioeconomic factors, and year of birth-adjusted odds of CHD without chromosomal abnormalities [adjusted odds ratio (OR) 1.4, 95\% confidence interval (CI) 1.1–1.7]. Assisted reproductive technologies were specifically associated with significant increases in the odds of malformations of the outflow tracts and ventriculoarterial connections (adjusted OR 1.7, 95\% CI 1.2–2.4) and of cardiac neural crest defects and double outlet right ventricle (adjusted OR 1.7, 95\% CI 1.1–2.7). In general, we found specific associations between methods of ART and subcategories of CHD.

Conclusion

Cases with CHD were more likely to have been conceived following ART when compared with malformed controls. This higher risk for CHD varied specifically according to the method of ART and the type of CHD and may be due to ART per se and/or the underlying infertility of couples.
Nuchal Translucency

- Measured at 11-14 weeks gestation.
- Cut-off values:
  - > 3 mm (95th centile): 15% CHD
  - > 3.5 mm (99th centile): 20% CHD
- Risk of CHD increases with NT measurement.
## Family History of CHD

<table>
<thead>
<tr>
<th>Family member affected</th>
<th>Risk</th>
</tr>
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<tbody>
<tr>
<td>Mother</td>
<td>3-7%</td>
</tr>
<tr>
<td>Father</td>
<td>2-3%</td>
</tr>
<tr>
<td>Sibling</td>
<td>3%</td>
</tr>
<tr>
<td>&gt; 1 sibling</td>
<td>6-12%</td>
</tr>
<tr>
<td>Second degree relative</td>
<td>2%</td>
</tr>
<tr>
<td>Third degree relative</td>
<td>1%</td>
</tr>
</tbody>
</table>
Timing of Fetal heart evaluation
MEDICAL TERMINATION OF PREGNANCY ACT

Medical termination of pregnancy Act, 1971 came into force from 1.4.1972. Implementing rule and regulations were revised in 1975, to simplify the procedure for approval of the place and for ready availability of services.

Conditions under which pregnancy can be terminated

Termination of pregnancy is permissible upto 20 weeks duration, if opinion is formed by one Registered Medical Practitioner (for pregnancies less than 12 weeks) or two registered medical practitioners (for 12-20 week preg.), in good faith, that:

1. Continuation of Pregnancy would involve a risk to the life of pregnant woman or grave injury to her physical or mental health.

2. There is a substantial risk of the Child born to suffer from such physical or mental abnormalities as to be seriously handicapped.

3. Pregnancy resulting from rape and from failure of contraceptive methods constitutes a grave injury to mental health of the woman. Account may also be taken of the woman’s actual or foreseeable environment in determining the risk to her health.
AIMS data: 2008-2014

Gestational age at referral (n=2451)
Integrating fetal heart screening with the anomaly scan:

Cost and time effective approach

Hunter S, Heads A, Wyllie J, Robson S

“Prenatal diagnosis of congenital heart disease in the Northern region of England: Benefits of a training programme for obstetric sonographers”

Heart 2000;84:294-298

Prenatal Diagnosis of Congenital Cardiac Anomalies: A Practical Approach Using Two Basic Views

Structural cardiac anomalies are estimated to occur in 8 of every 1,000 live births. Cardiovascular anomalies are frequently associated with other congenital anomalies because the heart is among the last organs to develop completely in the embryo. The guidelines for routine prenatal evaluation of both the American College of Radiology and the American Institute of Ultrasound in Medicine require evaluation of the fetal heart. The ultrasonographic (US) view that is most commonly used is the four-chamber view, which allows assessment of abnormalities involving the atria and the ventricles. However, this view is not adequate for demonstrating the outflow tracts of the aorta and pulmonary artery. A base view that demonstrates the crossing of the great vessels can be obtained just superior to the four-chamber view. Addition of the base view to routine US evaluation with the four-chamber view increases not only the sensitivity for detection of cardiac anomalies but also the accuracy of diagnosis.
Fetal Heart Screening: Cost effectiveness

- **Strategies analyzed:**
  - 4C, 4C + outflows, NC, Fetal echocardiography
  - 4C + Outflow tracts – least costly, effective
  - Fetal echo – most costly, effective
  - 4C+ Nuchal – costly, less effective

3-view technique

Unlikely to miss anything that is worth detecting.
Current expectations for adult survival after correction

Survival: 88.6% at 18 years

Moons et al
Circulation 2010;122:2264-2272
Impact of Fetal Diagnosis of Congenital Heart Defects: A projected Scenario for India

Critical Correctable Lesions
1. Planned delivery and neonatal care
2. In-utero therapy - medical/structural
3. Peri-partum cardiac teams
4. Improved outcomes
5. Financial support

Complex Lesions
• Better screening
• Early diagnosis (< 20 weeks)
• Multi system evaluation
• Option of TOP in relevant cases *
• Complex CHD burden reduce

Based on local, regional and national rules and regulations
Pregnancy outcomes in fetuses with CHD
AIMS data 2008-2015 ( n= 841)
PREGNANCY OUTCOMES IN FETAL CHD
Impact Of Type Of CHD

Impact of Type of CHD on Pregnancy Outcomes in Fetal CHD

- No live births
  - Simple: 32.7
  - Complex: 56.8

- Live births
  - Simple: 67.3
  - Complex: 43.2
PREGNANCY OUTCOMES IN FETAL CHD
Impact Of Gestational Age At Diagnosis

<table>
<thead>
<tr>
<th>Percentage</th>
<th>Non live births</th>
<th>Live births</th>
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<tbody>
<tr>
<td>77.4</td>
<td>22.6</td>
<td>32.2</td>
</tr>
<tr>
<td>67.8</td>
<td>32.2</td>
<td></td>
</tr>
<tr>
<td>77.2</td>
<td>22.8</td>
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Outcomes of Infants with Prenatally Diagnosed Congenital Heart Disease Delivered in a Tertiary-care Pediatric Cardiac Facility

Trupti Deepak Changlani, Annu Jose, Abish Sudhakar, Reshma Rojal, *Radhany Kunjikutty and Balu Vaidyanathan

Total N=121

Cardiac care n=96

NN Medical N=63
- NN Deaths N=6
- FU Alive * N=83
- FU Deaths N=5
- Lost FU N=2

NN Procedure N=33
- NN Alive N=33
- NN Deaths N=0
- FU Alive N=6
- FU Deaths N=0
- Lost FU N=8

Comfort care n=25

NN Deaths N=1

NN Survival N=14

* 13 Patient underwent procedures on FU
The Proposal

- Make Fetal heart evaluation mandatory component of all targeted anomaly scans
- Timing of scan: 16-20 weeks gestation
- Educate regarding the standard protocol for conduct and reporting of scan.
- Referral of cases which fail screening to higher level of expertise.
Action Plan

• Form a core advisory group – experts from fields of cardiology, Obstetrics, Radiology and Fetal Medicine.
• Training module and workshops – in Govt medical colleges/academic institutions/district hospitals
• Advanced training for selected doctors who wish for higher level of expertise (ToTs)
• Maintain a database of all records, follow-up of pregnancy and neonatal outcomes.
Training Modules: Format

- Duration: Half day.

- Awareness about CHD, indications for fetal echo.

- Technique of screening: lectures, Live demos, Hands-on training using locally available equipment.

- Educate on reporting fetal heart evaluation.

- Basic tips for counseling if anomaly detected

- Guidelines for referral.
Suggested downstream plan if an anomaly is detected …

- System for referral to the nearest pediatric cardiology expert.
- Counseling and pregnancy management planning.
- Option of planned delivery for correctable, critical problems.
- Support for heart surgery after birth.
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