Abstract

Congenital heart disease (CHD) is the most common congenital abnormality in human fetus, accounting for more than half of deaths from congenital abnormalities in childhood. Fetal echocardiography has been shown to be able to identify the majority of structural cardiac abnormalities, and it has traditionally been reserved for pregnancies at high risk for CHD. Most neonates with CHD, however, have no identifiable risk factors. When a sonogram is performed during pregnancy for defined clinical reasons, a four-chamber view of the fetal heart is routinely performed. However, a four-chamber view of the fetal heart does not reliably detect the most common CHD such as ventricular septal defect, coarctation of the aorta, transposition of the great arteries, and tetralogy of Fallot. Therefore, the vast majority of cases of CHD are left undetected even in those women who have undergone an obstetric ultrasound. A high level of suspicion of the presence of CHD and attention to anatomic details should be part of every ultrasound examination, especially when involving low-risk pregnancies, and it is currently recommended that only those fetuses with significant risk factors be referred for a targeted sonogram and fetal echocardiogram.

Keywords: Congenital heart disease; Fetal echocardiography; Four-chamber view
Intrauterine growth retardation
Fetal cardiac dysrhythmia
Fetal aneuploidy or other malformation
Polyhydramnios (AFI > 25)
Oligohydramnios (AFI < 5)
Abnormal four-chamber view, cardiac axis, or abnormal screening sonogram
Documented maternal viral or other infection known to affect fetal heart
Twin twin transfusion or multifetal gestation with discordance of fetal growth
Fetal macrosomia (estimated fetal weight of >4,500g) with evidence of cardiac compromise
Two vessel umbilical cord
Cardiac teratogen exposure
Before extensive fetal therapy such as fetal blood transfusion, fetal surgery
Marked abnormalities with Doppler interrogation of the fetal circulation
Decreased perfusion of vital organs during power Doppler evaluation or color flow mapping
Nonimmune hydrops fetalis

| 1. | Abnormal appearing heart on general fetal ultrasound examination |
| 2. | Maternal of family risk factors for cardiovascular disease, such as partent, sibling, or first (relative with congenital heart disease |
| 3. | Maternal diabetes mellitus (TGA, VSD, CA risk for fetus) |
| 4. | Maternal systemic lupus erythematosus (heart block risk for fetus) |
| 5. | Teratogen exposure during a vulnerable period |
| 6. | Other fetal organ system anomalies (including chromosomal) |
| 7. | Performance of transplacental therapy or presence of a history of significant but intermittent arrhythmia. Reevaluation examinations are required in these conditions |
| 8. | Fetal distress or dysfunction of unclear etiology |

1997 American Collage of Cardiology (ACC) American Heart Association (AHA)

| Intrauterine growth retardation |
| Fetal cardiac dysrhythmia |
| Fetal aneuploidy or other malformation |
| Polyhydramnios (AFI > 25) |
| Oligohydramnios (AFI < 5) |
| Abnormal four-chamber view, cardiac axis, or abnormal screening sonogram |
| Documented maternal viral or other infection known to affect fetal heart |
| Twin twin transfusion or multifetal gestation with discordance of fetal growth |
| Fetal macrosomia (estimated fetal weight of >4,500g) with evidence of cardiac compromise |
| Two vessel umbilical cord |
| Cardiac teratogen exposure |
| Before extensive fetal therapy such as fetal blood transfusion, fetal surgery |
| Marked abnormalities with Doppler interrogation of the fetal circulation |
| Decreased perfusion of vital organs during power Doppler evaluation or color flow mapping |
| Nonimmune hydrops fetalis |

(1) American Collage of Cardiology (ACC) American Heart Association (AHA)

| Intrauterine growth retardation |
| Fetal cardiac dysrhythmia |
| Fetal aneuploidy or other malformation |
| Polyhydramnios (AFI > 25) |
| Oligohydramnios (AFI < 5) |
| Abnormal four-chamber view, cardiac axis, or abnormal screening sonogram |
| Documented maternal viral or other infection known to affect fetal heart |
| Twin twin transfusion or multifetal gestation with discordance of fetal growth |
| Fetal macrosomia (estimated fetal weight of >4,500g) with evidence of cardiac compromise |
| Two vessel umbilical cord |
| Cardiac teratogen exposure |
| Before extensive fetal therapy such as fetal blood transfusion, fetal surgery |
| Marked abnormalities with Doppler interrogation of the fetal circulation |
| Decreased perfusion of vital organs during power Doppler evaluation or color flow mapping |
| Nonimmune hydrops fetalis |

10\% (combined ventricular output) |

(5) American Collage of Cardiology (ACC) American Heart Association (AHA)

| Intrauterine growth retardation |
| Fetal cardiac dysrhythmia |
| Fetal aneuploidy or other malformation |
| Polyhydramnios (AFI > 25) |
| Oligohydramnios (AFI < 5) |
| Abnormal four-chamber view, cardiac axis, or abnormal screening sonogram |
| Documented maternal viral or other infection known to affect fetal heart |
| Twin twin transfusion or multifetal gestation with discordance of fetal growth |
| Fetal macrosomia (estimated fetal weight of >4,500g) with evidence of cardiac compromise |
| Two vessel umbilical cord |
| Cardiac teratogen exposure |
| Before extensive fetal therapy such as fetal blood transfusion, fetal surgery |
| Marked abnormalities with Doppler interrogation of the fetal circulation |
| Decreased perfusion of vital organs during power Doppler evaluation or color flow mapping |
| Nonimmune hydrops fetalis |

10\% (combined ventricular output) |

(1) American Collage of Cardiology (ACC) American Heart Association (AHA)

| Intrauterine growth retardation |
| Fetal cardiac dysrhythmia |
| Fetal aneuploidy or other malformation |
| Polyhydramnios (AFI > 25) |
| Oligohydramnios (AFI < 5) |
| Abnormal four-chamber view, cardiac axis, or abnormal screening sonogram |
| Documented maternal viral or other infection known to affect fetal heart |
| Twin twin transfusion or multifetal gestation with discordance of fetal growth |
| Fetal macrosomia (estimated fetal weight of >4,500g) with evidence of cardiac compromise |
| Two vessel umbilical cord |
| Cardiac teratogen exposure |
| Before extensive fetal therapy such as fetal blood transfusion, fetal surgery |
| Marked abnormalities with Doppler interrogation of the fetal circulation |
| Decreased perfusion of vital organs during power Doppler evaluation or color flow mapping |
| Nonimmune hydrops fetalis |

10\% (combined ventricular output) |

(1) American Collage of Cardiology (ACC) American Heart Association (AHA)
Continuing Education Column

3. देश की इन्सुलिन की नष्ट होने से जुड़े दिब्लिटिस
Collagen vascular disease
Viral, bacterial, parasitic, or other infection known to affect fetal
or maternal heart
Rubella (PPAS, PDA, VSD, ASD risk for the fetus)a
Toxoplasmosis
Coxsackie virus
Cytomegalovirus
Mumps virus
Drug or teratogen exposure known to affect fetal heart
Lithium
Amphetamines
Alcohol
Anticonvulsant
Phenytoin
Trimethadione
Isoretinoin
Heavy metal toxicity
Maternal congenital or hereditary heart disease
Severe renal dysfunction uncorrected by dialysis or renal transplant
Advanced maternal age refusing chorionic villus sampling,
genetic amniocentesis, or triple screening
Phenylketonuria (tetrology of Fallot, ventricular septal defect,
atrial septal defect risk for the fetus)

* aPPAS, peripheral pulmonary artery stenosis; PDA, patent ductus
arteriosis; VSD, ventricular septal defect; ASD, atrial septal defect

Insulin-dependent diabetes mellitus
Collagen vascular disease
Viral, bacterial, parasitic, or other infection known to affect fetal
or maternal heart
Rubella (PPAS, PDA, VSD, ASD risk for the fetus)
Toxoplasmosis
Coxsackie virus
Cytomegalovirus
Mumps virus
Drug or teratogen exposure known to affect fetal heart
Lithium
Amphetamines
Alcohol
Anticonvulsant
Phenytoin
Trimethadione
Isoretinoin
Heavy metal toxicity
Maternal congenital or hereditary heart disease
Severe renal dysfunction uncorrected by dialysis or renal transplant
Advanced maternal age refusing chorionic villus sampling,
genetic amniocentesis, or triple screening
Phenylketonuria (tetrology of Fallot, ventricular septal defect,
atrial septal defect risk for the fetus)

Insulin-dependent diabetes mellitus
Collagen vascular disease
Viral, bacterial, parasitic, or other infection known to affect fetal
or maternal heart
Rubella (PPAS, PDA, VSD, ASD risk for the fetus)
Toxoplasmosis
Coxsackie virus
Cytomegalovirus
Mumps virus
Drug or teratogen exposure known to affect fetal heart
Lithium
Amphetamines
Alcohol
Anticonvulsant
Phenytoin
Trimethadione
Isoretinoin
Heavy metal toxicity
Maternal congenital or hereditary heart disease
Severe renal dysfunction uncorrected by dialysis or renal transplant
Advanced maternal age refusing chorionic villus sampling,
genetic amniocentesis, or triple screening
Phenylketonuria (tetrology of Fallot, ventricular septal defect,
atrial septal defect risk for the fetus)

10~12
18~20
28~30
20
### 4. Incidence of CHD

| Population         | (%) | Cardiac Defects<sup>a</sup> |
|--------------------|-----|-----------------------------|
| Normal Karyotype   | 0.8 | VSD, PDA, ASD               |
| Trisomy 22         | 65  | ASD, VSD, PDA               |
| Trisomy 21         | 50  | ECD, VSD, ASD, PDA          |
| Trisomy 18         | 99  | VSD, DORV, PS               |
| Trisomy 13         | 90  | VSD, PDA, Dext              |
| Trisomy 8          | 50  | VSD, ASD, PDA               |
| Trisomy 9          | 50  | VSD, CA, DORV               |
| 4p[]               | 40  | VSD, ASD, PDA               |
| 5p[]               | 25  | VSD, PDA, ASD               |
| 13q[]              | 25  | VSD                         |
| 14q[]              | 50  | PDA, ASD, Tet               |
| 18q[]              | 50  | VSD                         |
| 45x                | 35  | CA, AS, ASD                 |
| XXXXY              | 14  | PDA, ASD, ARC               |
| Triplody           | 50  | VSD                         |
| Cat[] eye syndrome | 40  | TAPVR, VSD, ASD             |

<sup>a</sup>VSD, ventricular septal defect; PDA, patent ductus arteriosus; ASD, atrial septal defect; TAPVR, total anomalous pulmonary venous return; Dext, dextrocardia; DORV, double outlet right ventricle; PS, pulmonary valve stenosis; ECD, endocardial cushion defect; CA, coarctation of the aorta; AS, aortic valve stenosis; ARC, anomalous right coronary artery; Tet, tetralogy of Fallot

### 3. Ultrasound

1. **Incidence of CHD**

2. **Left arm**

### Table

| Population         | (%) | Cardiac Defects<sup>a</sup> |
|--------------------|-----|-----------------------------|
| Normal Karyotype   | 0.8 | VSD, PDA, ASD               |
| Trisomy 22         | 65  | ASD, VSD, PDA               |
| Trisomy 21         | 50  | ECD, VSD, ASD, PDA          |
| Trisomy 18         | 99  | VSD, DORV, PS               |
| Trisomy 13         | 90  | VSD, PDA, Dext              |
| Trisomy 8          | 50  | VSD, ASD, PDA               |
| Trisomy 9          | 50  | VSD, CA, DORV               |
| 4p[]               | 40  | VSD, ASD, PDA               |
| 5p[]               | 25  | VSD, PDA, ASD               |
| 13q[]              | 25  | VSD                         |
| 14q[]              | 50  | PDA, ASD, Tet               |
| 18q[]              | 50  | VSD                         |
| 45x                | 35  | CA, AS, ASD                 |
| XXXXY              | 14  | PDA, ASD, ARC               |
| Triplody           | 50  | VSD                         |
| Cat[] eye syndrome | 40  | TAPVR, VSD, ASD             |

<sup>a</sup>VSD, ventricular septal defect; PDA, patent ductus arteriosus; ASD, atrial septal defect; TAPVR, total anomalous pulmonary venous return; Dext, dextrocardia; DORV, double outlet right ventricle; PS, pulmonary valve stenosis; ECD, endocardial cushion defect; CA, coarctation of the aorta; AS, aortic valve stenosis; ARC, anomalous right coronary artery; Tet, tetralogy of Fallot
Continuing Education Column

(Ebstein's anomaly), (pulmonary stenosis), (coarctation of aorta), Fallot (tetralogy of Fallot) (10, 11).

2.  
1)  
2)  

3.  (Ventricular Outflow Tracts)

1)  (Long axis View)
2) **Short axis View**

1. Use a **transducer** at 90° to capture the cardiac structures.

2. Use a **transducer** at 90° to capture the cardiac structures.

3) **Sagittal Views of the Great vessels, Aorta & Ductal Arches**

1. Use a **transducer** to capture the aortic arch.

2. Use a **transducer** to capture the aortic arch.

3. Use a **transducer** to capture the aortic arch.

4. Use a **transducer** to capture the aortic arch.
Continuing Education Column

7) “candy cane” (13)

4) “hockey stick” (9, 12).

1) (14–16).
2) 영문사전상, transducer에 대한 내용을 설명하고 있습니다.

3) Doppler를 이용한 혈류측정 방법을 설명하고 있습니다.

4) echo dropout을 이용한 방임의 방법을 설명하고 있습니다.

5) frequency transducer를 사용한 혈관조영술 방법을 설명하고 있습니다.
### 5. नुहन्त "करिमीय निर्माण अवस्था"

1. Is the cardiac apex to the left?
2. Are the right and left atrial volumes approximately equal?
3. Are the right and left ventricular volumes approximately equal?
4. Do the AV valves open equally widely?
5. Is the tricuspid valve annulus (right AV valve) displaced apically beyond the mitral annulus?
6. Is the moderator band identified in the apex of the right ventricle?
7. Is there a break in the ventricular septum or the atrial septum (other than the foramen ovale)?
8. Are the right and left ventricular free walls approximately the same thickness and are they normal for gestational age?
9. Are the mitral and tricuspid inflow velocities laminar by color Doppler and normal in velocity and configuration?
10. Is left ventricular contractility normal?

### 6. नुहन्त "करिमीय स्थान अवस्था"

- Hypoplastic left heart syndrome
- Hypoplastic right heart syndrome
- Atrioventricular canal defect
- Single ventricle
- Large ventricular septal defect
- Large atrial septal defect
- Valve atresia/stenosis
- Ebstein’s anomaly
- Double outlet right ventricle
- Moderate/severe coarctation of the aorta
- Cardiac tumors (rhabdomyomas)
- Cardiac situs abnormalities

### 7. नुहन्त "करिमीय स्थान अवस्था"

1. Are there two separate great vessels?
2. Does the anterior great vessel arise from the RV and course posteriorly, then bifurcate?
3. Does the posterior great vessel arise from the LV and course superiorly?
4. Do the great vessels ‘cross’ at the base?
5. Are the great vessels of approximately the same size?
6. Is color Doppler flow in each great vessel laminar and of normal velocity?
11. Hofman JI, Christian R. Congenital heart disease in a cohort of 19,502 births with long term follow up. Am J Cardiol 1978; 42: 641-7

12. Lian ZH, Zach MM, Erickson JD. Paternal age and occurrence of birth defects. Am J Hum Genet 1986; 39: 648-60

13. Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Circulation 1971; 43: 323-32

14. Ewigman BG, Crane JP, Frigoletto FD, LeFevre ML, Bain RP, McNellis D. Effect of prenatal ultrasound screening on perinatal outcome. RADIUS Study Group. N Engl J Med 1993; 329: 821-7

15. Cullen S, Sharland GK, Allan LD, Sullivan ID. Potential impact of population screening for prenatal diagnosis of congenital

---

**Diagram 1:**
- A: Aortic valve
- B: Right atrium
- B: Ventricles
- RA: Right atrium
- LA: Left atrium
- RV: Right ventricle
- LV: Left ventricle
- SVC: Superior vena cava
- IVC: Inferior vena cava
- MPA: Main pulmonary artery
- LPA: Left pulmonary artery
- PA: Pulmonary artery
- RPA: Right pulmonary artery
- AO: Aorta
- AO: Ascending aorta
- AO: Descending aorta
- AO: Aortic arch
- AO: Aortic isthmus
- AO: Aortic root
- AO: Aortic valve
- AO: Aortic annulus

**Diagram 2:**
- A: Aortic valve
- B: Right atrium
- B: Ventricles
- RA: Right atrium
- LA: Left atrium
- RV: Right ventricle
- LV: Left ventricle
- SVC: Superior vena cava
- IVC: Inferior vena cava
- MPA: Main pulmonary artery
- LPA: Left pulmonary artery
- PA: Pulmonary artery
- RPA: Right pulmonary artery
- AO: Aorta
- AO: Ascending aorta
- AO: Descending aorta
- AO: Aortic arch
- AO: Aortic isthmus
- AO: Aortic root
- AO: Aortic valve
- AO: Aortic annulus

---

**Diagram 3:**
- A: Aortic valve
- B: Right atrium
- B: Ventricles
- RA: Right atrium
- LA: Left atrium
- RV: Right ventricle
- LV: Left ventricle
- SVC: Superior vena cava
- IVC: Inferior vena cava
- MPA: Main pulmonary artery
- LPA: Left pulmonary artery
- PA: Pulmonary artery
- RPA: Right pulmonary artery
- AO: Aorta
- AO: Ascending aorta
- AO: Descending aorta
- AO: Aortic arch
- AO: Aortic isthmus
- AO: Aortic root
- AO: Aortic valve
- AO: Aortic annulus
Continuing Education Column

heart disease. Arch Dis Child 1992; 67: 775 - 8
6. Chitlin MD, Alpert J S, Armstrong WF, Aurigemma GP, Beller GA, Ryan TJ, et al. ACC/AHA guidelines for the clinical application of echocardiography. Executive summary. J Am Coll Cardiol 1997; 29: 862 - 79
7. Hess DB, Flaker G, Aggarwal KB, Buchheit LC, Hess LW. Fetal cardiac imaging. In: Hess DB, Hess LW, editors. Fetal echocardiography. Stamford: Appleton & Lange, 1999: 149 - 94
8. Yagel S, Weissman A, Rotstein Z, manor M, Hegesh J, Achiron R, at al. Congenital heart defects: natural course and in utero development. Circulation 1997; 96: 550 - 5
9. Abuhamad A. A practical guide to fetal echocardiography. Philadelphia: Lippincott[] Raven, 1997
10. Comstock CH. Normal fetal heart axis and position. Obstet Gynecol 1987; 70, 255 - 9
11. Shipp TD, Bromely B, Horberger LK, Nadel A, Benacerraf BR. Levorotation of the fetal cardiac axis: a clue for the presence of congenital heart disease. Obstet Gynecol 1995; 85: 97 - 102
12. Truesdell SC. Fetal cardiology. In: Jaffe R, Bui TH, ed. Textbook of fetal ultrasound. New York: Parthenon Publishing Group, 1999: 153 - 173
13. Friedman AH, Copel JA, Kleinman CS. Fetal echocardiography and fetal cardiology: Indications, diagnosis and management. Seminars in perinatology 1993; 17: 76 - 88