ORIGINAL RESEARCH PAPER

Radio Diagnosis

KEY WORDS: Prenatal diagnostic ultrasound, Congenital cardiac anomalies, Tetralogy of Fallot and Ebstein anomaly.

# A STUDY ON THE ANTENATAL SONOGRAPHIC EVALUATION OF CONGENITAL CARDIAC ANOMALIES IN PREGNANT WOMEN FOR A PERIOD OF TWO YEARS

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**ABSTRACT**

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 ultrasonographic (US) view like four-chamber view,three-vessel view, a base view, RVOT, and LVOT views are done during the fetal scan to detect cardiac abnormalities

**Technique**

In evaluating the fetal heart, the presentation and lie of the fetus should first be documented by obtaining longitudinal images of the cervix and uterine fundus. Following this, transverse images are obtained to determine the orientation of the fetal right side and left side. With a fetus in the supine or prone position, the spine becomes the point of reference in determining fetal orientation. If the fetus is lying on one side, a note is made as to whether the right or left side is dependent. The usual scanning sequence begins with the long axis of the thoracic spine; the transducer is rotated 90° until a four-chamber view of the heart is seen. Once the four-chamber view is visualized, the base view of the heart can be obtained by angling the transducer slightly cephalad or minimally sliding the transducer superiorly. It may be difficult to obtain high-quality images of the heart in a fetus with congenital heart disease. This is because other organs may be involved in a spectrum of anomalies.

**NormalViews**

The four chambers of the heart are delineated by the ventricular septum, the atrial septum, the tricuspid valve, and the mitral valve. The axis of the ventricular septum is directed to the left and forms an angle of 45°–50° with the midsagittal plane. The cardiac circumference is approximately one-third to one-half of the thoracic circumference throughout gestation. The foramen ovale interrupts the interatrial septum, with the flap of the foramen always opening toward the left atrium. The right ventricle is located behind the sternum and is characterized by the presence of the moderator band. Normally, both ventricles are approximately the same size. The left ventricle is posterior and to the left of the right ventricle, and the mitral valve insertion is slightly cephalad to the insertion of the tricuspid valve. These features help distinguish the right ventricle from the left ventricle.

The base view of the heart demonstrates the normal crossing of the great vessels just above the four-chamber view. The pulmonary artery arises from the right ventricle, proceeds across the midline, and passes anterior to the ascending aorta. As the pulmonary artery bifurcates, the right pulmonary artery loops around the circular ascending aorta. At this plane of bifurcation, the left pulmonary artery is not imaged. Instead, the ductus arteriosus forms the other limb of the bifurcation, connecting with the descending aorta. The pitfalls associated with this view lie primarily in the inability to obtain an optimum plane of scanning or failure to maintain the proper orientation as the fetus changes position. False-negative and false-positive diagnoses arise when both the aorta and the pulmonary artery are not seen in the same image. Assumptions should not be made as to the presence or normalcy of the vessel not imaged unless it is directly viewed. Also, constant reorientation of the transducer should be achieved by checking the position of the fetal head when a change in the fetal position is suspected.

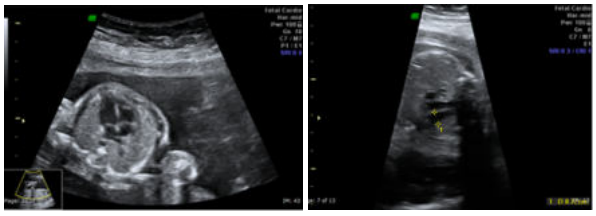

**Fig 1: four chamber view showing both ventricles, interventricular sepatum ,both atria ,interatrial septum and atrioventricular septum. Fig 2 : three vessel view showing from anterior to posterior we see pulmonary artery ,aorta and SVC.**

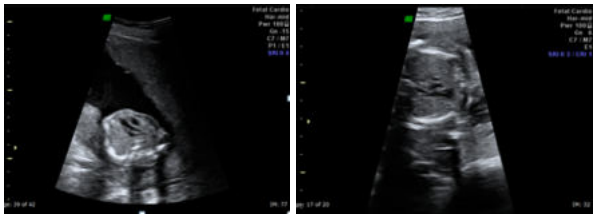

**Fig 3: LVOT showing the left ventricluar outflow tract arising from the left ventricle is seen clearly. Fig 4: RVOT showing the right ventricluar outflow tract arising from the right ventricle is seen clearly.**

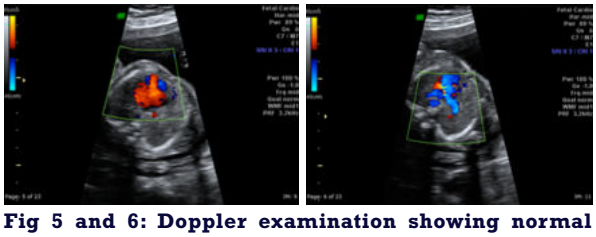

**Fig 5 and 6: Doppler examination showing normal ventricles and RVOT**

In this paper, we present 30 cases of congenital cardiac malformations in the rural population.

Out of 30 cases of congenital cardiac malformations in the rural area,1 case is of Ebstein anomaly, 2 case of cardiomegaly, 1 case of Aortic Stenosis, 1 case of hypertrophic cardiomyopathy, 2 cases of Tetrology of Fallot, 4 case of Hypoplastic left heart syndrome, 1 case of Hypoplastic right heart syndrome, 7 cases of ventricular septal defects, 3 cases

of echogenic intracardiac foci, 2 cases of aberrant right subclavian artery, 1 case of Persistent left SVC, 1 case of the common arterial trunk 3 case of cardiac axis deviation and 2 case of pericardial effusion were found.

| | N = 30 |
|---|---|
| **TYPE OF CARDIAC MALFORMATION** | **NO OF CASES** |
| Ebstein anomaly | 1 |
| Cardiomegaly | 2 |
| Aortic Stenosis | 1 |
| Hypertrophic cardiomyopathy | 1 |
| Tetrology of Fallot | 2 |
| Hypoplastic left heart syndrome | 4 |
| Hypoplastic right heart syndrome | 1 |
| Ventricular septal defect | 7 |
| Echogenic intracardiac foci | 3 |
| Aberrant right subclavian artery | 1 |
| Persistent left Superior vena cava | 1 |
| Common arterial trunk | 1 |
| Cardiac axis deviation | 3 |
| Pericardial effusion | 2 |

**Ebstein anomaly**

Ebstein anomaly is characterized by displacement and attachment of one or more tricuspid leaflets toward the apex of the RV. It accounts for less than 1% of congenital heart defects and occurs in 1 per 20,000 live births. It is associated with maternal lithium use, chromosomal abnormalities, ASD, patent foramen ovale, and pulmonary stenosis or atresia. Ultrasound shows the apical displacement of the tricuspid valve into the RV, tethered leaflets, reduction in the size of the functional RV, and tricuspid regurgitation. Intrauterine mortality is as high as 85%. Differential diagnosis includes tricuspid valve dysplasia and idiopathic RA enlargement.

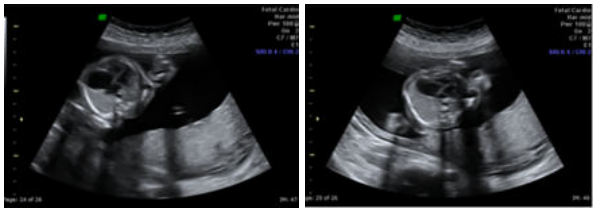

**Fig 7 and 8 : Antenatal sacn showing dilated right atrium.**

**Aortic Stenosis**

Aortic stenosis is narrowing of the LVOT that can be seen at the valvular, supra-valvular, or subvalvular level, with an incidence of 5 % in newborns. Valvular stenosis may be associated with chromosomal abnormalities and bicuspid aortic valve, subvalvular stenosis with hypertrophic cardiomyopathy or inherited disorders, and supra-valvular stenosis with William syndrome. Ultrasound diagnosis is difficult and may show thickening of the aortic valve leaflets, with hypertrophied LV and dilated aorta as a result of post stenotic dilation. Doppler ultrasound shows high velocities across the valve (Fig. 16) and helps differentiate it from atresia.

In the below images (Fig 7 and 8 )we can clearly see the decreased calibre of aorta near its origin with increased PSV of aorta near the stenosis ( > 130 cm/sec )

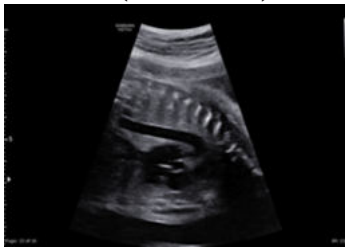

**Fig 9 : Showing decreased calibre of aorta near its origin**

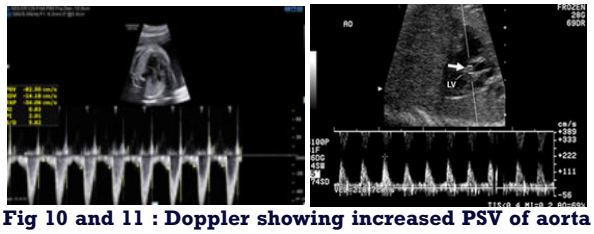

**Fig 10 and 11 : Doppler showing increased PSV of aorta near the stenosis**

**Hypertrophic cardiomyopathy**

Cardiomyopathies account for 8–11% of fatal cardiovascular abnormalities, with one-third of fetuses dying in utero. Cardiomyopathies can be broadly classified as dilated, hypertrophic, and restrictive types. Intrinsic causes of primary cardiomyopathy are single-gene disorders (Noonan syndrome, familial cardiomyopathy, and metabolic abnormalities), mitochondrial and storage disorders, chromosomal abnormalities, and αthalassemia. Extrinsic causes are intrauterine infections, maternal diseases (autoantibodies and diabetes), and twin-twin transfusion syndrome. In hypertrophic cardiomyopathy, the LV-RV myocardial thickness is increased, without an underlying structural abnormality. It has been associated with maternal diabetes and often regresses during the first six months of life.

In the below image (Fig 12), we can clearly see the increased thickness of the right and left ventricular walls.

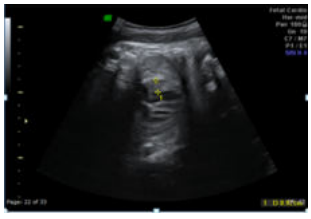

**Fig 12: showing increased thickness of the right and left ventricular walls**

**Tetrology of Fallot**

Tetralogy of Fallot is characterized by narrowing of the RVOT, VSD, overriding aorta, and right ventricular hypertrophy. It accounts for 5–10% of congenital cardiac defects and is seen in 1 per 1000 live births. The incomplete closure of the septum results in aortic overriding. It is associated with chromosomal and extracardiac abnormalities. On ultrasound, the aorta is seen straddling a large membranous VSD. The aorta may be dilated, and the pulmonary valve is stenosed or atretic with a dilated PA. Because of the presence of normal fetal shunts, RV hypertrophy is not seen in the fetus.

In the below images (Fig 13, 14 and 15 )we can clearly see the components of TOF like: overriding of aorta, VSD and Pulmonic stenosis.

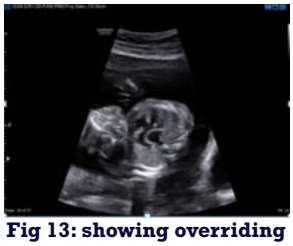

**Fig 13: showing overriding of aorta**

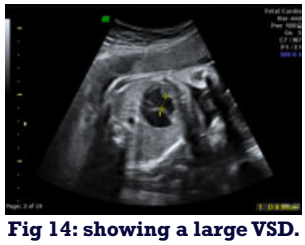

**Fig 14: showing a large VSD.**

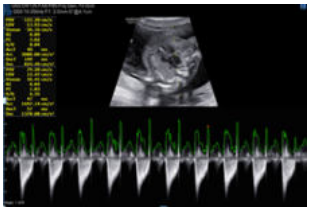

**Fig 15: Doppler showing increased PSV of pulmonary artery.**

**Hypoplastic left heart syndrome**

Hypoplastic left heart syndrome is characterized by hypoplastic left-sided cardiac structures, including the LV, mitral valve, aortic valve, and aorta. It accounts for 2 % of congenital cardiac defects and is seen in 1 per 3000 live births. It is more common in boys and is caused by decreased flow in and out of the LV during development (e.g., mitral or aortic stenosis or atresia). Blood flow to the systemic circulation (coronary arteries, brain, liver, and kidneys) in these patients is dependent on flow through the ductus arteriosus. It is associated with aortic coarctation in 80% of cases. On ultrasound, the LV is small (LV: RV ratio < 1) in size; the ventricular septum makes an angle of 90° with the spino-sternal line, and the aortic outflow is smaller than the pulmonary outflow tract. Mitral and aortic valves are hypoplastic or atretic. A single area of flow is seen at the AV level and bidirectional flow at the proximal aorta because of distal aortic coarctation.

In the below images (Fig 16 and 17) we can see the decreased volume of the left ventricle.

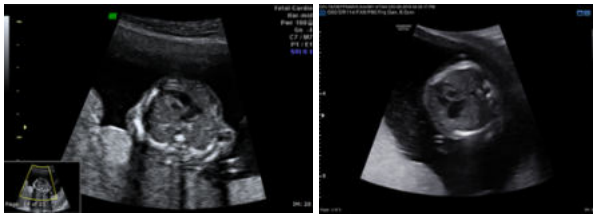

**Fig 16 and 17: showing decreased volume of the left ventricle.**

**Ventricular septal defect**

Ventricular septal defect (VSD) is the most common congenital heart disease, seen in 3 per 1000 live births, and accounting for 30% of all cardiac anomalies. The defect is most commonly (80%) seen in the membranous septum and less commonly in the muscular, outlet, or inlet portions. VSD is best seen in a four-chamber view as a discontinuity in the ventricular septum, particularly the inlet defects. Small defects can be difficult to detect, particularly in the perimembranous portion, but Doppler imaging can show flow across the defect. In isolated VSD, bidirectional shunting with right-to-left shunt during systole and left-to-right shunting in diastole is seen, but in VSD associated with other anomalies, unidirectional shunting may be seen. Small defects may close, but large defects require surgical closure.

In the below images (Fig 18 and 19) we can see the membranous and muscular VSD.

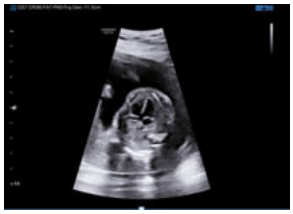

**Fig18: showing membranous VSD**

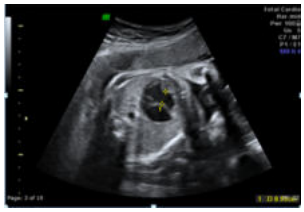

**Fig 19: showing membranous and muscular VSD**

**Cardiomegaly**

Fetal cardiomegaly (FC) refers to an enlarged fetal heart. It is variably defined as a fetal cardio-thoracic circumference above two standard deviations. Fetal cardiothoracic (C/T) circumference ratio is the ratio of the cardiac circumference to the thoracic circumference and may be easily measured on fetal ultrasound. It can arise from a number of situations like tricuspid atresia, Ebstein anomaly, cardiomyopathies, etc.

In the below images (Fig 20 )we can see the enlarged cardia with increased cardiothoracic ratio

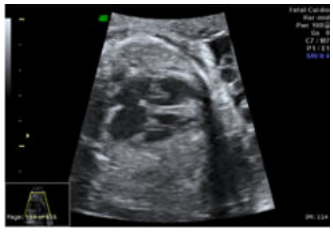

**Fig 20 : Antenatal sacn showing enlarged cardia with increased cardiothoracic ratio**

**Echogenic intracardiac foci**

Echogenic intracardiac foci are seen in 3 % of fetal hearts, more commonly in the LV than the RV representing the reflection of the ultrasound waves of the small papillary muscle or chorda tendinae. It is usually insignificant but may be associated with chromosomal anomalies, such as trisomy 13 or 21, where papillary muscles or chordae may be calcified. Detection of echogenic intracardiac foci increases the likelihood of Down syndrome. Calcification can also be seen in cardiac neoplasms, but neoplastic calcifications are larger, multiple, and not as echogenic as the echogenic intracardiac foci.

In the below images (Fig 21 and 22 )we can see the single and double intracardiac echogenic foci in the left ventricle

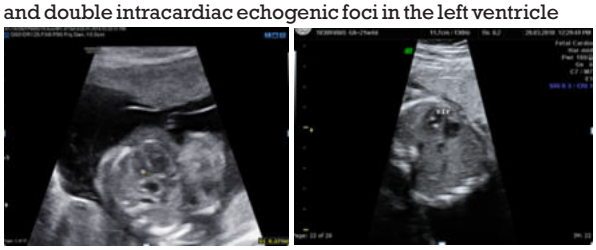

**Fig 21 and 22: showing single and double intracardiac echogenic foci.**

**Aberrant right subclavian artery.**

Aberrant right subclavian arteries (ARSA), also known as arteria lusoria, are one of the commonest of the aortic arch anomalies. Instead of being the first branch (with the right common carotid as the brachiocephalic artery), it arises on its own as the fourth branch, distal to the left subclavian artery. It then hooks back to reach the right side with its relationship to the esophagus variable:80% posterior to the esophagus, 15% between esophagus and trachea,5% anterior to the trachea.

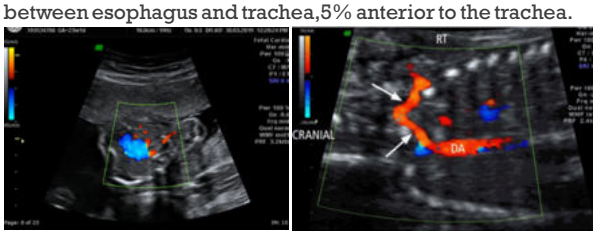


**Fig 23 and 24 : Doppler examination showing aberrent right subclavian artery arisisng separately from aorta.**

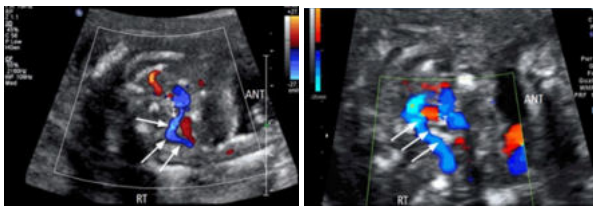


**Fig 25 and 26: Dopplor examination showing aberrent right subclavian infront of trachea ( Normal ) and behind the trachea which is abnormal.**

**Persistent left Superior vena cava**

A left-sided superior vena cava (SVC) is the most common

congenital venous anomaly in the chest, and in a minority of cases, can result in a right-to-left shunt. A left-sided SVC is seen in 5% of those with congenital heart disease. A left-sided SVC forms when the left anterior cardinal vein is not obliterated during normal fetal development. The persistent left-sided SVC passes anterior to the left hilum and lateral to the aortic arch before rejoining the circulatory system. There are a number of possible drainage sites. One is coronary sinus, and the other is the left atrium.

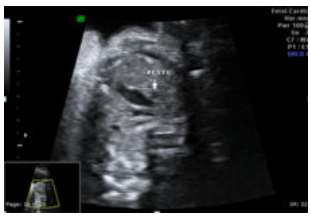

**Fig 27: showing SVC on left side.**

**Common arterial trunk**

Truncus arteriosus is characterized by a single arterial trunk that feeds the systemic pulmonary circulation and coronary arteries with a single semilunar valve. It accounts for 1–2% of congenital cardiac defects, is seen in 0.08–0.16 per 1000 live births [8], and is caused by the failure of fusion and descent of the conotruncal ridge. It almost always straddles a VSD and receives blood from both the ventricles but rarely originates almost completely from the RV or LV. There are four types (Collett Edwards classification) based on the level of origin of the aorta and pulmonary arteries [9]. An admixture of oxygenated and deoxygenated blood in the common trunk results in subnormal systemic oxygenation. The ductus arteriosus is not necessary for systemic flow and, therefore, does not fully develop. On ultrasound, a single arterial trunk is seen overriding the interventricular septum, with an associated VSD, and there are several branches connecting with the aorta and pulmonary vasculature.

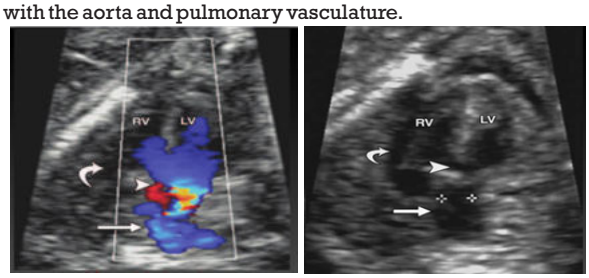


**Fig 28 and 29 : Dopplex examination showing common origin of single vascular trunk from the ventricles with associated VSD**

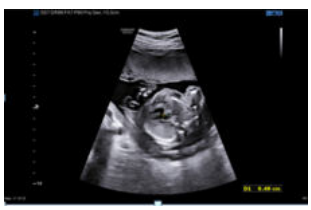

**Fig 30: VSD in patient with trunkus arteriosus.**

**Cardiac axis deviation**

The cardiac axis is the angle the interventricular septum makes with the anteroposterior diameter of the thorax. The normal cardiac axis is 45± 15°. The heart normally deviates to the left. It is almost entirely within the left chest. An altered cardiac axis is a pointer toward an anomaly. An altered axis is often associated with outflow tract anomalies.

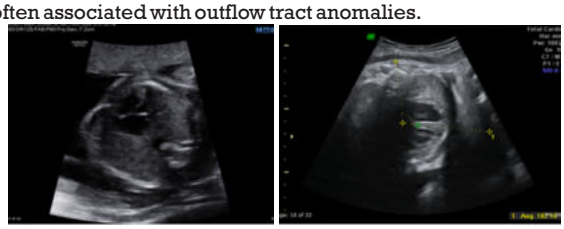

**Fig 31 and 32 : Showing altered cardiac axis.**

**Pericardial effusion**

Fetal pericardial effusions occur when there is an accumulation of pericardial fluid in utero. In order to be considered abnormal, it is generally accepted that the pericardial fluid thickness should be greater than 2 mm. A fetal pericardial effusion can occur as a component of hydrops fetalis: where it is usually one of the earliest findings in hydrops and other cardiac abnormalities like arrhythmias and fetal cardiac tumors.

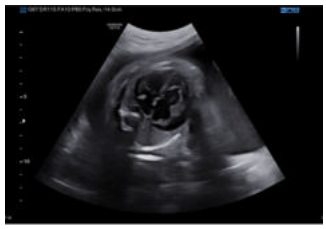

**Fig 33 : Showing fluid in the pericardial cavity.**

## CONCLUSION

Routine fetal cardiac ultrasound using four-chamber and outflow-tract views enables the detection and characterization of most of the cardiac anomalies. A further comprehensive evaluation can be performed with fetal echocardiography, particularly in high-risk pregnancies and extracardiac anomalies. Doppler imaging is used in the evaluation of vascular and valvular lesions.

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