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Review Article

What is NOT normal in fetal heart

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ABSTRACT

Congenital heart defects are leading cause of neonatal mortality even in developed world. It is result of either late diagnosis of defect after circulatory compromise or discharge of neonates with undiagnosed defect. Antenatal anomaly scan provides an important opportunity to suspect congenital heart defects. Various approaches have been proposed by international society for ultrasound in obstetrics and gynecology (ISUOG), American heart association (AHA) etc to screen fetal heart in various views. In spite of best of practices, not less than 30 to 40% of congenital heart diseases are born without antenatal suspicion of same. Steep learning curve associated with understanding of cardiac anatomy is one of the most important reason. During antenatal scan, important step is to suspect cardiac abnormality. We wish to stress upon 'What is NOT normal ?' in afore mentioned protocols to arouse suspicion. This can initiates early refer for fetal echo and parent counseling by pediatric cardiologist as well as planned delivery in tertiary setup with paediatric cardiac backup. Early referral is also important where laws about medical termination of pregnancy are restrictive and time bound.

1. Introduction

Incidence of Congenital Heart Disease (CHD) is 8 to 9 per 1000 live births as per the reported data worldwide. Out of which 1.6 to 2.6 per 1000 live births are the critical congenital heart diseases which are particularly at risk of hemodynamic compromise and mortality in early neonatal period. Even in first world countries , more than third of these neonates are discharged from nursery without suspicion of heart disease and these undiagnosed babies are at highest risk of morbidity and mortality. This makes congenital heart disease leading cause of infant mortality.[1–4] Antenatal ultrasound (US) examination plays crucial role in early diagnosis of CHD's. Antenatal diagnosis rate of CHD's remains around 60 to 70% world wide. Significant number of CHDs remain undiagnosed antenatally because of lack of operator skills and knowledge. As it in not possible to include detail fetal echocardiogram in routine antenatal US examination, various protocols have been proposed (e.g basic cardiac echocardiac examination BCEE). Now fetal cardiac evaluation include extended cardiac echocardiographic examination, BCEE + outflow tract view (OTV)+ 3 vessel trachea view (3VTV). With increasing range of examination, sensitivity has increased from 50 % (BCEE) to 83% (BCEE+ OTV+ 3VTV).[5–12]
Most important step in antenatal scan to diagnose a heart defect is to suspect deviation from normal. Current article stresses on "what is NOT normal?" and simple clues to suspect the same. These can be helpful to the operator who is not extensively trained in imaging of fetal heart. This can help to initiate early referral to the tertiary care centers for detail fetal echo cardiography, parent counseling by pediatric cardiologist. Timely referral is also important where medical termination of pregnancy is under tight legal scrutiny.

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2. Examination Protocol

We like to offer to some tips to simplify image acquisition and interpretation. Details of it can be found in relevant text.

1. Use cardiac set up of machine in gray scale with curvilinear probe or sector probe. Individual preferences are important. Some features like continuous wave Doppler are available with Sector probe.
2. Ideal fetal position may not be possible but chest anterior is most preferable.
3. Take a caudo-cranial swipe from foetal abdomen to the base of neck to look for situs, position, and axis of heart and for any major deviation from normal. Replay it in slow motion. It help to understand orientation of various structures with one another.

First standard normal views are discussed. What is not normal or what can arouse suspicion of abnormality in a said view has been discussed along with differential diagnosis. Description of individual lesion is out of preview of this topic and it can be found in relevant text.

4 chamber view: (Figures 1, 2, 3, 4, 5 and 6

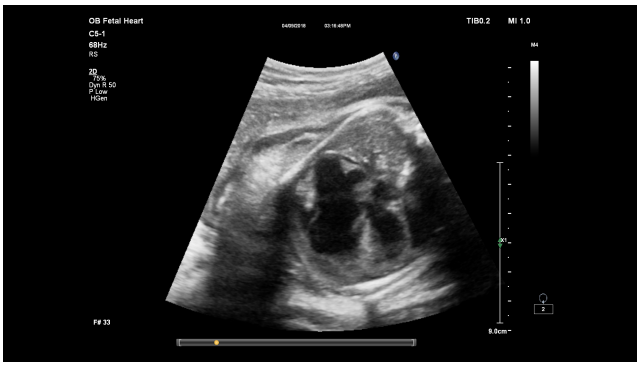

Fig. 1: Small left ventricle reaching up to apex

This is one of the most easily obtained view and can offer very valuable information. We can categories it into following broad subtypes. It is particularly more relevant during second trimester.

1. **All 4 chambers are seen and no discrepancies in size of ventricle** i.e. normal 4 chamber view: This is seen in a normal heart and conotruncal anomalies like tetralogy of Falllot (TOF), trans position of great arteries (TGA), double outlet right ventricle (DORV), truncus arteriosus. It effectively rules out Hypoplastic left heart syndrome (HLHS), Tricuspid atresia (TA), single ventricle etc.[2,7,10]
2. **Both ventricles are reaching upt o apex but one looks smaller than other**: If left ventricle (LV) is small, possibilities are coarctation of aorta, aortic arch interruption, total anomalous pulmonary venous connection, hypoplastic mitral valve, congenital mitral anomalies. If right ventricle (RV) is small, pulmonary atresia intact ventricular septum (PAIVS), hypoplastic right heart syndrome, pulmonary stenosis are possibilities.[13–16]
3. **There is a dominant ventricle and other ventricle is very small and not reaching up to apex**: Small, non apex forming ventricle will have hypoplastic atrioventricular valve. In most cases of HLHS, LV does not reach upto apex and may be dysfunction along with endocardial fibroelastosis. With non apex forming RV, TA and PA IVS are possible.[13,14,17]
4. **Only one ventricle is appreciated and other ventricle not seen**. The dominant ventricle can be LV, RV or indeterminate morphology. There may be one or two AV valves draining atrium or atria into the ventricle. These are univentricular hearts. If there is a very large ventricular septal (VSD) defect, it can also

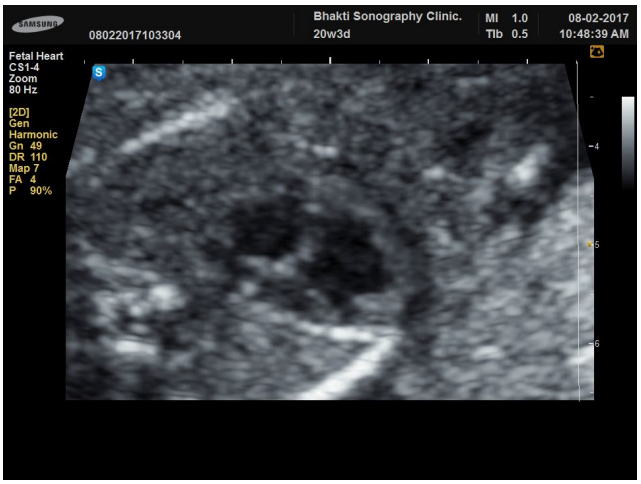

Fig. 2: Small Left ventricle not reaching up to apex

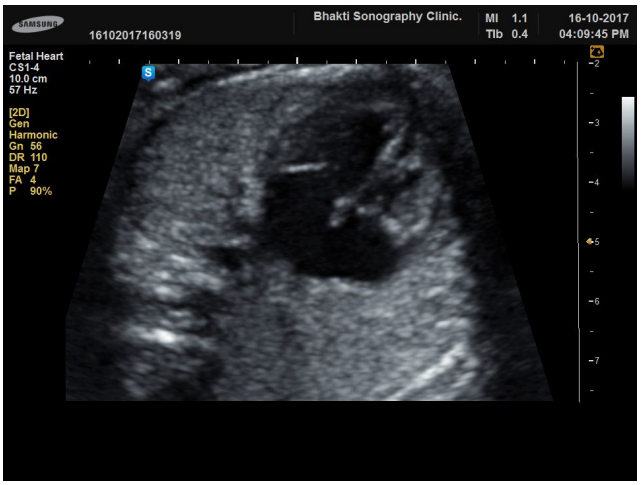

Fig. 3: Small Right ventricle reaching up to apex

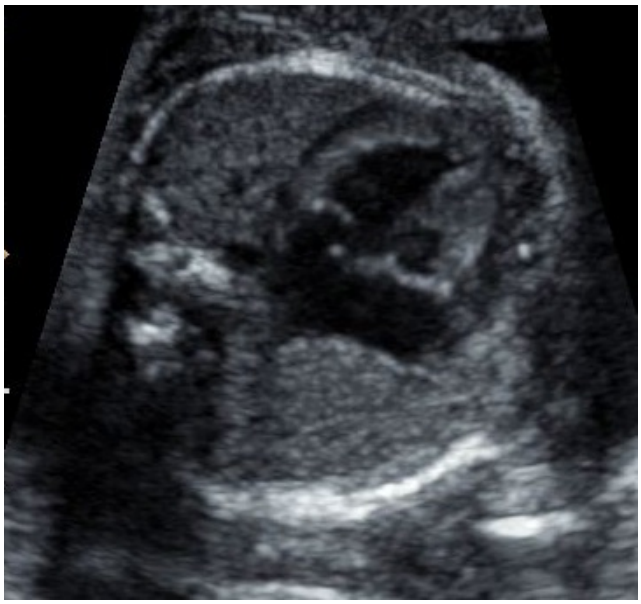

Fig. 4: Small Right ventricle not reaching up to apex

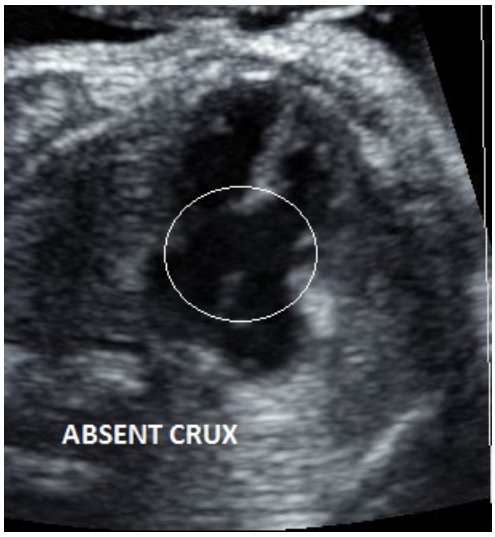

Fig. 5: Complete atreiventricular septal defect: note common AV valve

give impression of single ventricle.[18]

5. **Abnormally dilated ventricle**: Mitral or tricuspid valve regurgitation and systolic dysfunction of ventricle has to be sought for. LV dilatation and dysfunction can result from severe aortic stenosis, endocardial fibroelastosis, cardiomyopathy etc. RV dilatation or dysfunction can result from severe tricuspid regurgitation in Ebstein anomaly or PAIVS, cardiomyopathy etc. Fetal tachycardia (supra ventricular tachycardia, atrial flutter) and bradycardia (complete atrioventricular block) can also cause ventricular dysfunction and hydrops fetalis.[14,17,19–21]

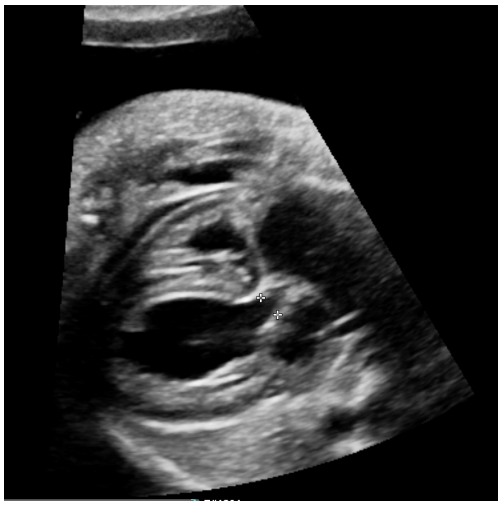

Fig. 6: Dilated LV, note endocardial fibroelastosis in a case of severe aortic stenosis

6. **Flow in oval foramen**: Normally flow in oval foramen is from right to left with septum primum bulging into left atrium. In cases of HLHS, there is flow reversal. If foramen is very small with restrictive flow, outcome of HLHS is very poor.[14]

In a heart with complex anomalies, it may not be possible to determine morphology of ventricle and AV valve but describing them as they are seen may be more informative.

Outlet: (Figures 7 and 8)

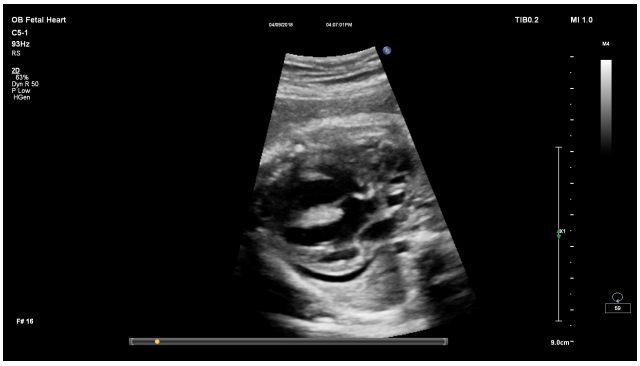

Fig. 7: Mal aligned VSD with single outlet and other outlet can not be traced in a Tetralogy of Fallot and pulmonary atresia

Ventricular outlet can be obtained 1) tilting probe cranially from 4 chamber view or 2) short axis view. Following points if present should arouse suspicion of CCHD

1. **Semilunar valve over riding VSD/ Mal aligned VSD/ Double outlet ventricle**: In a normal heart and defects like TGA/ congenitally corrected trans position of great arteries (CCTGA) with intact septum,

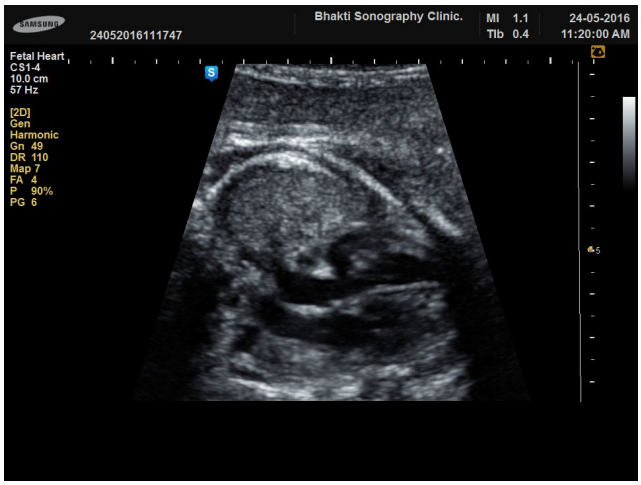

Fig. 8: Paralleloutlet and great vessels

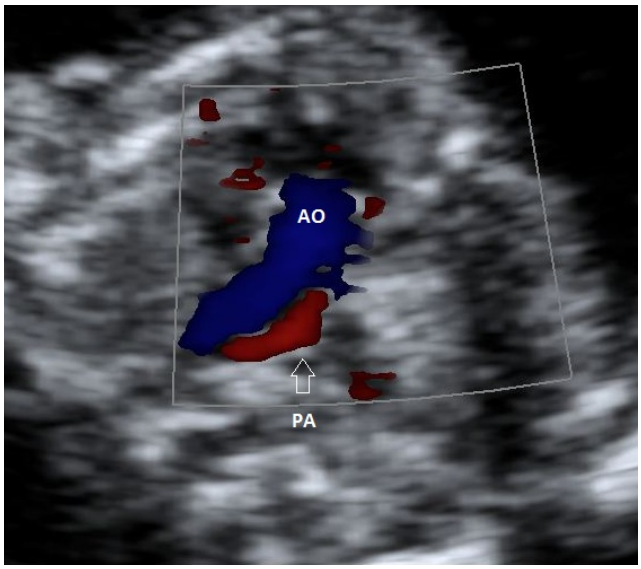

Fig. 9: Dominant aorta and small pulmonary artery with flow reversal

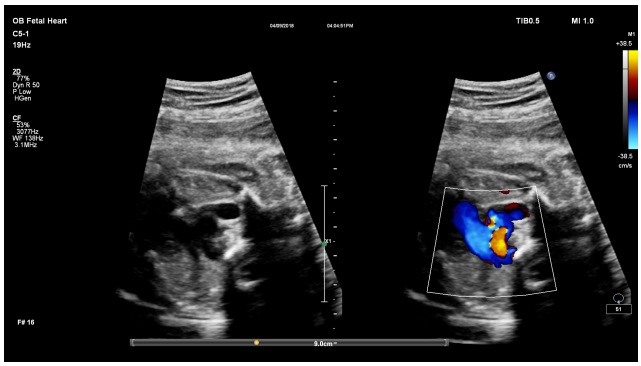

Fig. 10: Dominant pulmonary artery and small aorta with flow reversal

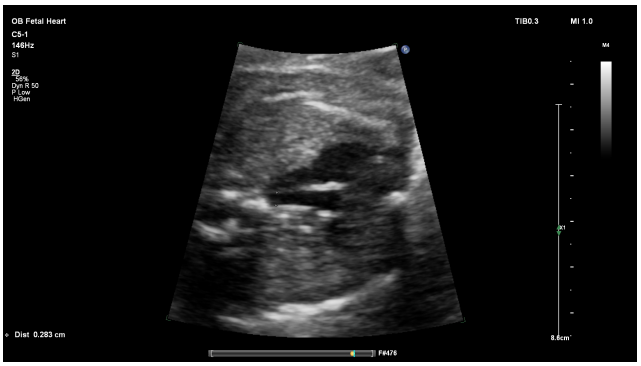

Fig. 11: Dominant pulmonary artery and small aorta

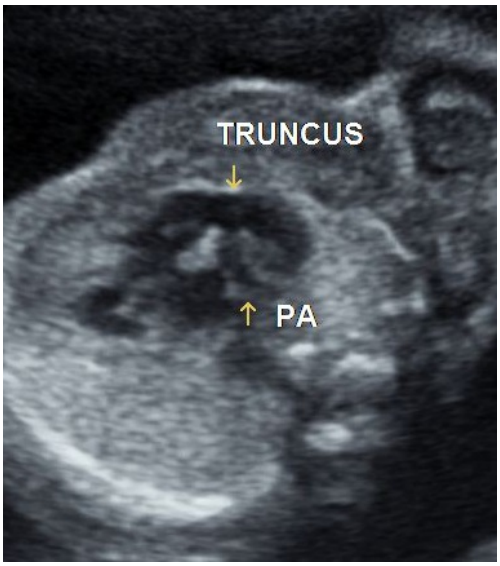

Fig. 12: Single great vessel arising from ventricle

one semilunar valve (aortic/ pulmonary) is committed to each ventricle. Same is true even with isolated perimembranous or outlet VSD. If an imaginary line is drawn from inter-ventricular septum, annulus of a semilunar valve is completely on one side of it and other semilunar valve on other side. However, if a semilunar valve is seen arising from both ventricles with a VSD underneath it, it is called as semilunar valve over riding VSD/ mal aligned VSD. It is a very unique feature seen with many conotruncal defects like TOF with or without pulmonary atresia, TGA VSD, CCTGA VSD, truncus arteriosus etc as well as isolated mal aligned VSD. Occasionally single ventricle can have mal aligned outlet foramen (VSD). DORV is an unique condition where both semilunar valves are committed to right ventricle. This arrangement can be seen with uni ventricular hearts like tricuspid atresia also. Very rarely both outlet arises from left ventricle.[22,23]

2. **Crossing out let vs parallel outlet:** In normal heart outlet of left and right ventricle are crossing to each other. In short axis they give appearance of cup and saucer. This arrangement can also be seen with TOF and a few cases of DORV. However, TGA, CCTGA, DORV are characterized by parallel arrangement of outlets and great vessel. It may be difficult to determine morphology of individual outlet, but presence of parallel vessel is a tell tale sign on complex anomaly.[22,23]
3. **Outlet/Semilunar valve:** Presence of flow acceleration, small and thick valve are indicator of valve stenosis but assessment of severity may not be easy. Presence of only one semilunar valve may indicate atresia of other valve or truncus arteriosus. Mal aligned VSD with only one semilunar valve is seen in setting of TOF/ TGA/ DORV/ CCTGA with pulmonary atresia, truncus arteriosus or aortic atresia with VSD. Single ventricles can also have pulmonary or aortic atresia. Exact delineation of morphology of patent semilunar valve may be very challenging and assessment of morphology of great vessels is very useful.[14–16,22,23]
4. **Great vessels (3 vessel view)**: (Figures 9, 10, 11 and 12) This view can be obtained by moving cranially from 4 chamber view with a sweep**.** In a normal heart superior vena cava, aorta and main pulmonary artery/ arterial duct are arranged from Rt to Lt. This arrangement is disturbed in most conotruncal anomalies except TOF and a few DORV. Vessels are arranged in a triangular fashion in TGA. Aorta is left and anterior in CCTGA. However, as mentioned earlier for a examiner, who is not very accustomed to fetal cardiac anatomy, any deviation from normal should be considered as warning sign.[14–16,22,23] Sizes and relative proportion of vessel are important anatomical findings. During second trimester, pulmonary artery/ aorta ratio is up to 1.2/1. In case of coarctation, pulmonary artery is dilated while aorta may have distal tapering. A ratio more than 2: 1 is highly suggestive of same. Coarctation of aorta can be associated with VSD, TGA VSD, DORV, TA with transposition of great arteries, HLHS, single ventricle. In may of these conditions, neonatal emergency result of critical coarctation leading to shock and not the associated cardiac defect. Small pulmonary artery is seen with critical pulmonary stenosis in isolation or with TOF, and other conotruncal anomalies.[14–16,22,23] Single vessel in outlet chamber can be seen with HLHS, pulmonary atresia either isolated or with another anomalies, truncus arteriosus etc. In aortic atresia, aorta can be very small, and it may not be appreciated on plain ultrasound. However, color flow and Doppler assessment can show flow reversal in distal aortic arch. Same is possible with pulmonary atresia, where flow reversal in arterial duct can be appreciated. In case of TOF pulmonary atresia with major aorto pulmonary collateral, flow reversal in arterial duct can not be appreciated as it is not formed. While, pulmonary artery in truncus arteriosus can be tricky to appreciate.[14–16,22,23]

Few simple tricks that are followed to report cardiac defect during anomaly scan.

1. Individual view can not be interpreted in isolation. Always correlate with other segments. Take a sweep from caudal end to cranial end and run it in a loop at a slower speed. It helps to analyze anatomy better.
2. Color Doppler for flow assessment.
3. If one segment is not seen, then may not be necessarily absent, it can be just very small.
4. Use simple anatomical terms. Cardiac anatomy at times can be very confusing hence it is better to describe what one sees than to use anatomical jargon.

3. Conclusion

Cardiac examination is an integral part of antenatal anomaly scan. However, complexity of congenital heart defect makes detail assessment difficult for an inexperienced examiner. To suspect an abnormality, one need not know entire spectrum cardiac morphology. Understanding of deviation from normal and keeping high index of suspicion for same can arouse suspicion and early referral. All these aspects are important for better care of neonates with CHD and appropriate parent counselling and informed decision making. .

4. Source of funding

None.

5. Conflict of interest

None.

References

1. Botto LD, Correa A, Erickson JD. Racial and temporal variations in the prevalence of heart defects. *Pediatr*. 2001;107:32.
2. Mahle WT, Newburger JW, Matherne GP, Smith FC, Hoke TR, et al. American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Interdisciplinary Council on Quality of Care and Outcomes Research, American Academy of Pediatrics Section on Cardiology, Cardiac Surgery. Committee on Fetus and Newborn. Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. *Pediatr*. 2009;124(2):823–836. Available from: 10.1542/peds.2009-1397.
3. Brown KL, Ridout DA, Hoskote A, Verhulst L, Ricci M, Bull C. Delayed diagnosis of congenital heart disease worsens preoperative condition and outcome of surgery in neonates. *Heart*. 2006;92:1298–1302.

4. Kemper A, Mahle W, Martin G, Cooley WC, Kumar W, et al. Strategies for Implementing Screening for Critical Congenital Heart Disease. *Pediatr*. 2011;128(5).
5. Velzen CLV, Clur SA, Rijlaarsdam M, Bax CJ, Pajkrt E, et al. Prenatal detection of congenital heart disease-results of a national screening programme. *BJOG*. 2016;123:400–407.
6. Quartermain MD, Pasquali SK, Hill KD, Goldberg DJ, Huhta JC, et al. Variation in Prenatal Diagnosis of Congenital Heart Disease in Infants. *Pediatr*. 2015;136(2):378–385. Available from: e378-e385.doi:10.1542/peds.2014-3783.
7. Carvalho JS, Allan LD, Chaoui R, Copel JA, Devore GR, et al. ISUOG practice guidelines (updated): sonographic screening examination of the fetal heart. *Ultrasound Obstet Gynecol*. 2013;41:348–359.
8. Mrginean C, Tognel R, Gozar L, Muntean I, Mrginean CO, et al. -. *Obstetrics*. 2010;6:82–86.
9. Singh Y, Geoch LM. Fetal Anomaly Screening for Detection of Congenital Heart Defects. *J Neonatal Biol*. 2016;5:115. Available from: 10.4172/2167-0897.100e115.
10. Donofrio MT, Moon-Grady AJ, Hornberger LK, Copel JA, Sklansky MS, et al. Diagnosis and Treatment of Fetal Cardiac Disease A Scientific Statement From the American Heart Association. *Circ*. 2014;129:2183–2242.
11. Zhang MA, Zeng YF, Zhao XL, Lu EF, HW. Diagnostic Value of Fetal Echocardiography for Congenital Heart Disease A Systematic Review. *Med*;94(42):1759–1759.
12. Achiron R, Glaser J, Gelernter I. Extended fetal echocardiographic examination for detecting cardiac malformations in low risk pregnancies. *BMJ*. 1992;304:671–674.
13. Yagel H, Kivilevitch Z, Valsky DV, Achiron R, Yagel S, et al. The fetal venous system: normal embryology, anatomy, and physiology and the development and the appearances of anomalies. *Informa Healthcare*;10017:413–426. Fetal cardiology.
14. Allen LD. Left heart Malformation. vol. 10017. Yagel S, Silverman N, Gembruch U, editors. NY ;.
15. Yoo SJ, Bradly T, Jaeggi E. Aortic arch anomalies. vol. 10017. Yagel S, Silverman N, Gembruch U, et al., editors ;.
16. Fauron JC. Anomalies of right heart. vol. 10017. Yagel S, Silverman N, Gembruch U, editors. NY ;.
17. Carvelho JS, Yagel S, Silverman N, Gembruch U. Pulmonary atresia with intact ventricular septum. *Informa Healthcare*;10017:269–280. Fetal cardiology.
18. Chouvi R. Cardial malposition and syndrome with right and left isomerism. Yagel S, Silverman N, Gembruch U, et al., editors ;.
19. Simpson J. Cardiomyopathies in fetus. Yagel S, Silverman N, Gembruch U, et al., editors ;.
20. Schmidt K. Fetal Bradydysrhythmia. Yagel S, Silverman N, Gembruch U, et al., editors ;.
21. Gemburch U. Fetal Tachyarrhythmia. Yagel S, Gembruch U, Silverman N, editors ;.
22. Yoo SJ, Golding F, Jaeggi E. Ventricular outflow tract anomalies: so called conotruncal anomalies. vol. 10017. Yagel S, Silverman N, Gembruch U, et al., editors ;.
23. Fermont L. Developments in diagnosis of transposition of great arteries. Yagel S, Silverman N, Gembruch U, et al., editors ;.

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