CASE REPORT

Ventricular and Great Artery Disproportion during routine Fetal Heart Imaging Evaluation and Management / Technical Report

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ABSTRACT

Size discrepancy (disproportion) between left and right cardiac chambers (atria, ventricles) and /or great arteries is easily identified during routine mid-gestational ultrasound screening (basic fetal heart imaging), representing a referral indication for detailed fetal echocardiogram to rule out the presence of fetal congenital heart disease. In the present review the appropriate imaging technique to avoid foreshortening of ventricular chambers during basic fetal heart imaging, non-cardiac causes of heart chamber disproportion as well as common congenital heart defects associated with chamber and artery disproportion during routine ultrasound fetal imaging are presented.

KEY WORDS Fetal ovarian cyst; abdomen cyst; prenatal diagnosis

Clinical examples

Case 1. Great artery and ventricular disproportion with right ventricular and pulmonary artery dominance over left ventricle and aorta, respectively, was diagnosed during routine sonographic evaluation in a female fetus at 34 wks of gestation. After birth the neonate had signs of diminished femoral pulses, the clinical suspicion of aorta isthmus stenosis (coarctation) has been confirmed by echocardiography. Surgical treatment followed within the first month of life.

Case 2. Pronounced great artery and ventricular disproportion, with diminutive non-apex forming left ventricle, associated hypoplasia of mitral valve, aortic valve, aorta ascendens and transverse aortic arch, detected during routine "extended basic" fetal heart sonographic evaluation at mid-gestation (19weeks). Fetal caryotype was normal, while the diagnosis of hypoplastic left heart syndrome was confirmed following referral for fetal echocardiogram, family was informed about the guarded long-term prognosis and the need for three cardiosurgical procedures after birth followed by increased probability of heart transplantation later in life.

Case 3. Ventricular disproportion, with right ventricular dominance, mild disproportion of great arteries (pulmonary artery dominance) associated with a central septal wall defect (atrial and ventricular) in 4-chamber view and the presence of a common atrioventricular valve. Fetal echocardiogram followed confirming the diagnosis of unbalanced atrioventricular septal defect (AVSD), trisomy 21 has been detected following amniocentesis.

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Definition

Disproportion is defined as an obvious size difference when comparing two structures. The Greek terms dys-analogy (abnormal proportions) or a-symmetry (lack of symmetry) express also the visual size imbalance among structures. The evaluation of disproportion most commonly is subjective (qualitative), expressing the visual impression of the observer when comparing two structures. However for the confirmation of the subjective visual impression exact measurement of each structure size and estimation of the size ratio is needed (objective assessment)

The subjective evaluation for the presence –or absence-of symmetry between fetal heart structures is a fundamental part of the routine fetal heart imaging during mid-gestation sonographic evaluation of fetal anatomic surveillance. Due to the contribution of both fetal circulations to the combined cardiac output (connected at atria level with foramen ovale and arterial level with the ductus arteriosus), the left and right fetal heart structures are visually equally in size. The documentation of symmetry between left and right fetal heart structures is mandatory to be documented both for the atria and ventricles, evaluated at 4 chamber view during "basic" fetal heart imaging, as well as for the great arteries, evaluated at 3 vessel/ 3 vessel-trachea view during "extended" basic fetal heart imaging. The absence of symmetry between left and right fetal heart structures, when imaging errors are excluded, is indicative of imbalance in fetal heart loading and /or growth.

According to current guidelines for the performance of routine sonographic fetal heart evaluation (1) as an essential element of midgestation sonographic fetal anatomy surveillance, the sonographer should evaluate the integrity of fetal cardiovascular system, based on the detailed evaluation of five consecutive transverse views of fetal thorax, from upper abdomen included up to mediastinum. These include the 1) upper abdomen, 2) four chamber (4CH), 3) left ventricular outflow tract (LVOT), 4) Right ventricular outflow tract (RVOT), 5) 3 vessel view (3V) and 6) 3 vessel and trachea (3V-T) view.

Disproportion during routine fetal heart imaging

The presence of symmetry between left and right fetal heart chambers is an essential element of a normal four chamber view (Figure 1): 1) two atria, approximately equal in size, should be documented. They are differentiated as left and right atrium not only from their relative position (the left atrium being the most posterior fetal heart chamber, anterior to the spine, the right atrium being more anterior and rightward relative to the left atrium) but also due to distinct anatomic landmarks: pulmonary veins enter into the left atrium, while the foramen ovale flap projects into the left atrium. 2) two ventricles, approximately equal in size, should be also documented. Similarly they are differentiated as left and right ventricle not only based on their relative position (the right ventricle is the most anterior fetal heart chamber just behind the fetal sternum, the left ventricle posterior and leftward from the left ventricle, with the intraventricular septum having a normal angle of 45 degrees relative to the anteroposterior fetal thorax axis) but also due to distinct anatomic features: the right ventricle atrioventricular valve (tricuspid) offset is more close to the fetal apex compared to the left ventricle atrioventricular valve (mitral), the right ventricle has a muscular band close to its apex (moderator band). (1-4)

The presence of symmetry between the great arteries can be already assessed in their proximal parts, as they originate from the corresponding ventricles in a "cross-over" fashion (LVOT and RVOT views) as the proximal aorta has a rightward (towards right shoulder) and the proximal pulmonary artery a straight backward direction (towards left paravertebral area).

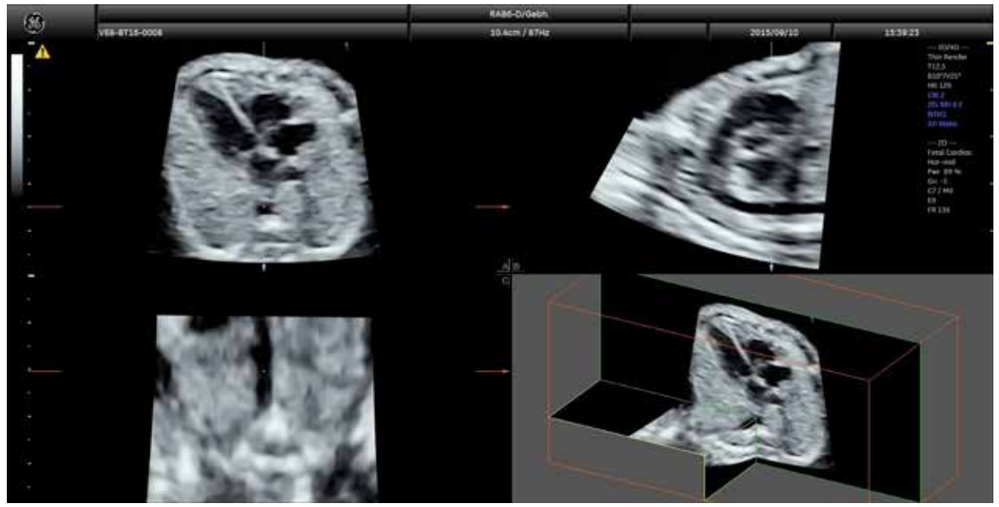

Figure 1. *Appropriate transverse plane for routine fetal heart imaging: 4 chamber view*
STIC volume reconstruction, demonstrating the recommended 4 chamber view transverse plane (upper left, original acquisition plane A), acquired by scanning the fetal thorax perpendicular to the fetal spine-aorta (upper right, reconstructed plane B), which are visualized at a large segment (lower left, reconstructed plane C). Author's personal archive

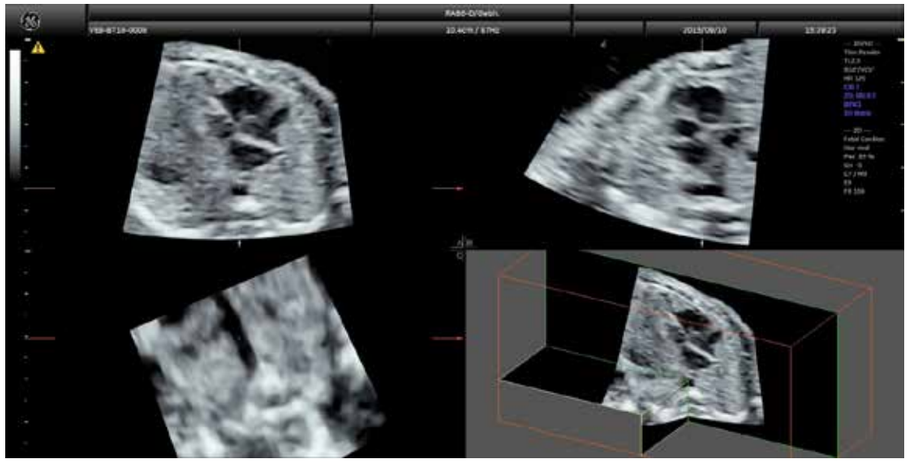

Figure 2. *Pseudo-disproportion due to misalignment of 4-Chamber view plane*
STIC volume reconstruction, demonstrating ventricular pseudo-disproportion (RV>LV) obtained by a non-perfect transverse fetal thorax view (upper left, original acquisition plane A), acquired by scanning the fetal thorax perpendicular to the fetal spine-aorta (upper right, reconstructed plane B), which are although not visualized at a large segment (lower left, reconstructed plane C) due to incomplete rotation of the probe to the transverse plane. Author's personal archive

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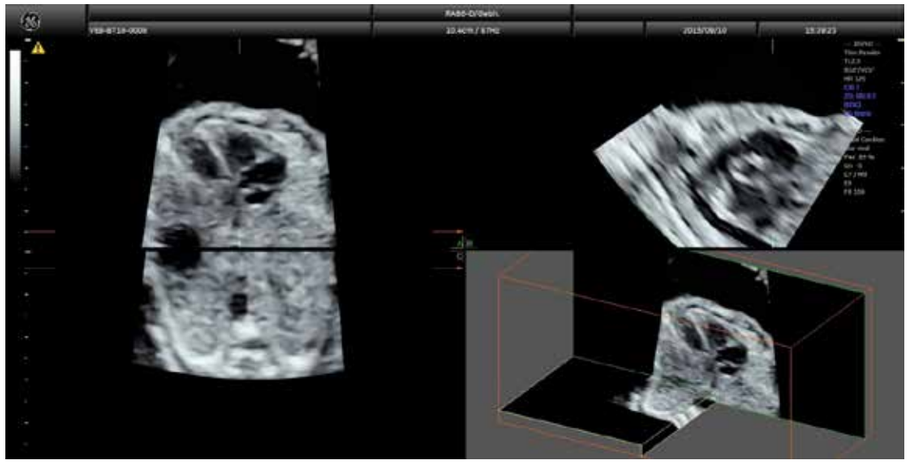

Figure 3. *Pseudo-disproportion due to misalignment of 4-Chamber view plane*
STIC volume reconstruction, demonstrating ventricular pseudo-disproportion (RV>LV) obtained by a non-perfect transverse fetal thorax view (upper left, original acquisition plane A), acquired by scanning the fetal thorax oblique to the fetal spine-aorta (upper right, reconstructed plane B), due to unfavorable fetal position. Author's personal archive

The assessment of symmetry in the distal parts of the great arteries (transverse aorta compared to distal pulmonary artery –ductus arteriosus) is evaluated at the more superior mediastinal views, namely at the 3 vessel view and 3 vessel-trachea view. In the 3 vessel view, from right to left are visualized the cross section of the right superior vena cava, the transverse aortic arch (with a leftward now direction), and the pulmonary artery – ductus arteriosus (straight backward direction). In the 3V trachea view, both great arteries (aorta and ductus arteriosus) confluence at the left of the trachea (named the V sign), where the aorta descendent accepts their combined flow. In the 3 vessel view a mild asymmetry of the vascular structures is expected representing a normal finding, with the superior vena cava being of smaller diameter compared to aorta, which is relative smaller than the pulmonary artery-ductus arteriosus. It should be emphasized that the presence of cross-over of the great arteries and a V sign at their confluence at the left of the trachea is not sufficient to identify the artery originating from the left ventricle as aorta, and the one originating of the right ventricle as the pulmonary artery: the distinct anatomic features of each vessel should be also demonstrated for their correct characterization (the pulmonary artery giving branches –bifurcation very close to its proximal part, the aorta's branches originate much more distally).

Pseudo-disproportion.

This term is suggested as most appropriate in the present review, to describe the visual impression of fetal heart chamber disproportion when such a disproportion does not really exist, a finding based solely due to imaging related limitations (Figures 2,3). Pseudo-disproportion is a common referral indication for fetal echocardiography, especially in cases where the fetal position is unfavorable, combining a twisting and bending fetal projection, or when there is a failure to obtain a perfect transverse cut of the fetal thorax, due to unfavorable fetal spine projection relative to any available ultrasound probe angle placed on the maternal abdominal area.

An ideal 4 chamber view should be obtained at a perfect transverse fetal thorax sonographic plane (Figure 1). First we have to evaluate whether the sonographic plane is appropriate (perfect transverse) and then to validate fetal heart chamber symmetry. The correct imaging plane should be perpendicular (at 90 degrees) to the axis of the fetal spine: This can be accomplished by first vis-

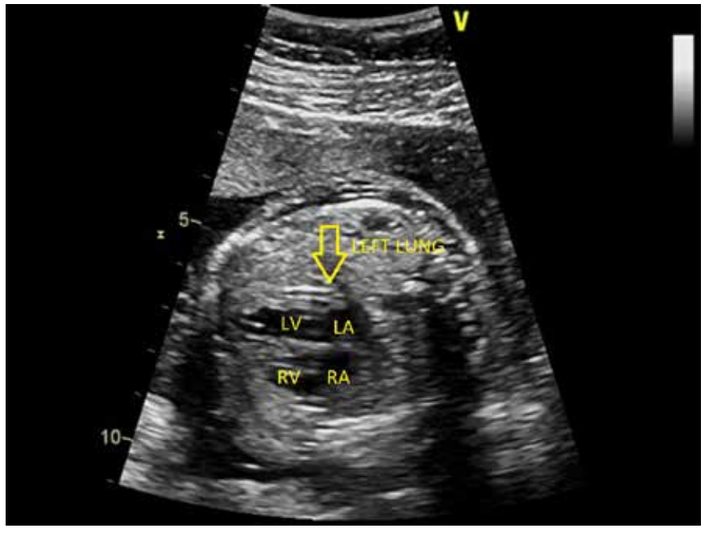

Figure 4. *Fetal Heart Disproportion due to external fetal heart compression*
External compression by left lung mass (CCAM) resulting into rightward displacement of fetal heart, abnormal heart axis and mild disproportion (LV<RV) of ventricles in 4 chamber view. Author's personal archive

ualizing the fetal spine in a sagittal view (where the full length of the thoracic spine is in view), first by moving / angling our probe to the maternal abdomen so as the fetal spine lies parallel to our probe interface, then carefully rotating the probe (while keeping the same insonation angle relative to spine) at 90 degrees, and adapting our transverse plane to the recommended fetal heart imaging plane by sliding the probe towards the fetal head or abdomen, as needed. In cases where the fetal projection is a combination of twist and bending it could be impossible to have a perfect transverse view, as indicated by the imaging of multiple rib cuts in the one side of the fetal thorax compared to the other. Care therefore should be taken to comment on the present of fetal heart chamber symmetry or not, when a perfect transverse imaging plane was not or cannot be acquired. Repeating the scan at a later point and referring the case for fetal echocardiography in case of persistent fetal chamber disproportion despite repeated imaging efforts, is recommended.

Non-cardiac causes of disproportion.

Fetal heart position, size and symmetry within the fetal thorax can be affected by non-cardiac conditions resulting in heart compression or translocation within the fetal thorax (Figure 4). The normal fetal heart imaging in 4 chamber view is characterized by a) both stomach and heart being at the left side of the fetus b) heart occupies one third or thoracic area (cardiothoracic area ratio < 0.33), c) the majority of fetal heart area lies in the left chest and d) the cardiac axis (apex) points to the left, with an angle of 45 degrees (± 20 degrees) between the anteroposterior thorax line (sternum to spine) and the line crossing the fetal intraventricular septum. In case of fetal heart chamber disproportion associated with any abnormalities of the above normal 4-chamber view features, including abnormal fetal heart position within the fetal thorax, abnormal fetal heart size or abnormal fetal heart axis care should be taken to rule out non-cardiac causes resulting into fetal heart compression including congenital diaphragmatic hernia, fetal lung malformations with mass effect on adjacent structures (lung sequestration, cystic adenomatous malformation, lung cysts), lung hypoplasia and fetal thorax deformities

Disproportion due to loading conditions.

Provided that pseudo-disproportion and non cardiac causes of disproportion have been excluded, differences in loading conditions between left and right fetal heart

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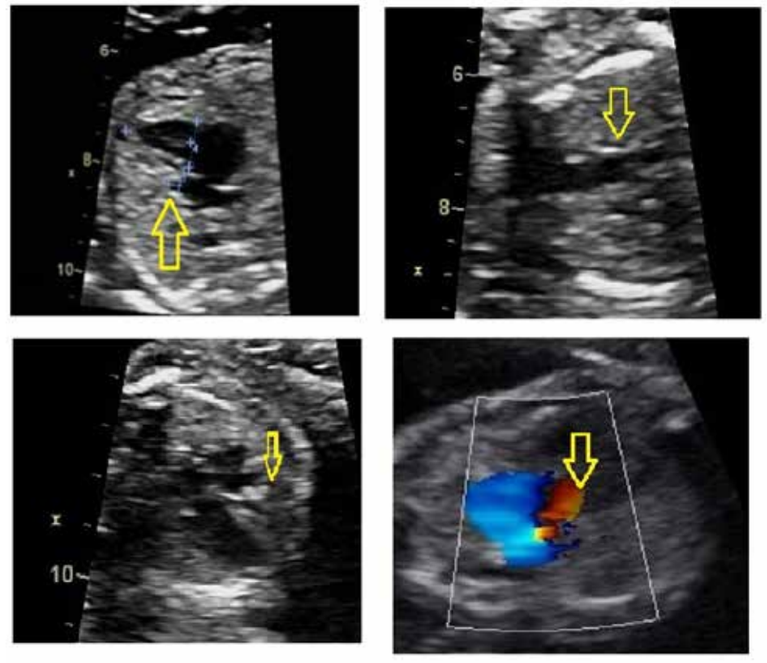

Figure 5. *Ventricular and Great Artery disproportion due to Critical Fetal Congenital Heart Disease*
Severe hypoplasia of left atrium-diminutive left ventricle (upper left) and aorta (upper right) in Hypoplastic Left Heart Syndrome; Hypoplasia and hypertrophy of right ventricle (lower left) with retrograde flow in hypoplastic pulmonary artery (lower right) in Pulmonary Atresia-Intact Ventricular Septum (arrows). Author's personal archive

chambers have to be also excluded. In contrast to postnatal circulation, where the two circulations (pulmonary, systemic) are connected in series (all systemic venous blood return will pass through the right heart chambers into pulmonary circulation, then to left heart chambers, systemic arteries and back to right heart), in the fetus the two circulations are connected in parallel (communicating in the atrial level-foramen ovale and arterial level-ductus arteriosus, both with right to left shunt) (5). On contrast to postnatal circulation where the left ventricle (LV) is the dominant pump, in the fetus the right ventricle (RV) is dominant, contributing about 60% of combined fetal cardiac output. RV filling is based mainly on inferior and inferior vena cava flow (deoxygenated venous blood) while it pumps relative deoxygenated blood through the ductus arteriosus (DA) into the fetal aorta descendens and placenta (fetal lungs receive very low perfusion). LV filling is based mainly on the highly oxygenated blood, of umbilical vein-ductus venosus (DV), entering preferentially into the left atrium through the foramen ovale intraatrial communication. LV pumps the highly oxygenated blood into aorta ascendens and fetal brain, the remaining passing through the narrowest part of the aorta (isthmus) to the aorta descendens (which receives also the flow from the ductus arteriosus). Different loading conditions (in terms of filling –preload and resistance against which the ventricles have to work-afterload) can result in ventricular disproportion, including cavity size and / or wall thickness. A typical example of ventricular disproportion due to differential loading conditions is ductus arteriosus (DA) constriction, associated with significant afterload increase for the RV, resulting into RV dilation and RV/LV disproportion. Careful inspection of DA flow pattern and size can allow for the appropriate diagnosis, as ventricular disproportion at late gestation is also associated with the possibility of congenital heart

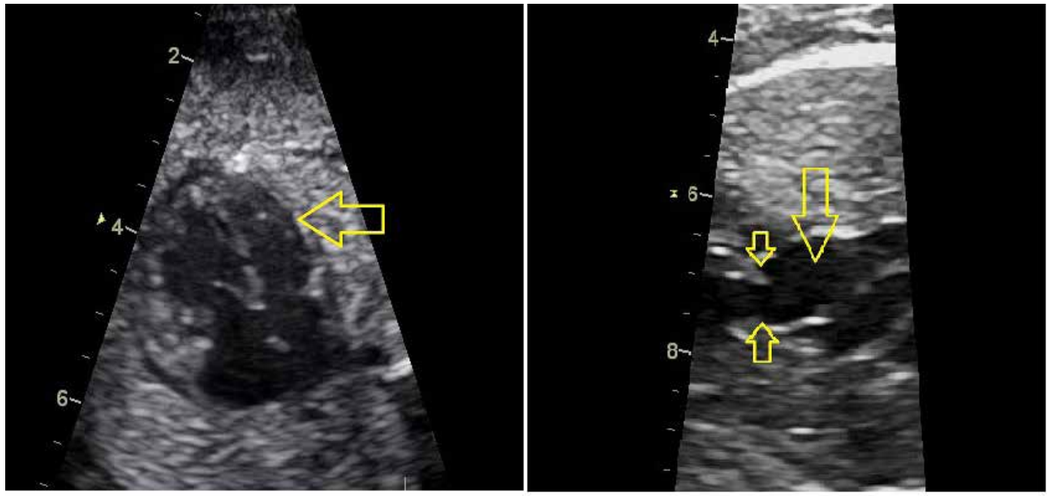

Figure 6. *Ventricular and Great Artery disproportion due to Mild-Moderate Fetal Congenital Heart Disease*
Mild Ventricular disproportion (LV <RV) in atrioventricular septal defect (AVSD) (left); Mild Great Artery disproportion (PA>AO) in valvar pulmonary stenosis associated with systolic doming of thickened pulmonary valve leaflets (smaller arrows) (right). Author's personal archive.

disease (especially aortic isthmus stenosis / coarctation of the aorta). Avoidance or discontinuation of administration of prostaglandin inhibitors (NSAIDs) to the pregnant woman with DA constriction can result in recovery of normal DA flow pattern and ventricular disproportion (6). Other causes of non-congenital heart disease associated ventricular disproportion include restrictive flow in the foramen ovale (in animal models resulting into hypoplastic left heart syndrome), increased placenta flow vascular resistance etc. Cases of fetal anemia or high flow states (hyperdynamic circulation associated with AV malformations) can lead to increased fetal heart size with relative balanced ventricular size.

Disproportion due to congenital heart disease.

Provided that pseudo-disproportion and fetal heart compression and loading imbalances have been excluded, the possibility that fetal heart artery chamber and /or great artery disproportion is due to fetal congenital heart disease (CHD) is very high (7). There is an immediate referral indication for fetal echocardiogram by an expert fetal cardiologist, with referral indication "abnormal fetal heart views during anomaly scan, suspected fetal congenital heart disease" (2-4). The detailed description of all CHD types associated with ventricular and /or great artery disproportion is out of the scope of this review, as CHD forms associated with disproportion are numerous with a great variability of the presence and extent of disproportion observed also within any given CHD form (for example tetralogy of Fallow can be associated with various degrees of pulmonary hypoplasia) as well as within the same subject with advancing gestational age (dynamic evolution of CHD forms during pregnancy) (5). As general rule, the more pronounced the disproportion and the earlier in gestation when the disproportion is detected, the more guarded the final prognosis of the fetus regarding the possibility of a final complete bi-ventricular repair might be. An example of extreme disproportion (RV> LV) with diminutive left heart structures (LV, Aorta) at times hardly to detect at all represents the hypoplastic left heart syndrome (HLHS, one of the most severe CHD forms) (Figure 5). Similar findings regarding the right ventricle can be observed in pulmonary atresia with intact ventricular septum, characterized by extreme hypertrophied, diminutive right ventricle (Figure 5). Ventricular and great artery disproportion can be observed in cases of aortic isthmus stenosis (coarctation) at times

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associated with long segment aortic arch hypoplasia (7). Cases of aortic and pulmonary artery valve stenosis, especially if not critical, can be associated with postenotic dilation of the corresponding artery, resulting in great artery disproportion (Figure 6). Atrial and ventricular disproportion is a common finding in complex congenital heart disease associated with malformation of atrioventricular valves (including tricuspid valve atresia with ventricular septum defect, Ebstein malformation of tricuspid valve characterized by a massively enlarged right atrium and some forms of atrioventricular septal defect –non balanced AVSD forms, Figure 6).

In every case of fetal heart chamber / artery disproportion during the routine mid-gestational anomaly scan, not explained due to fetal position, external fetal heart compression or loading imbalance, there is an immediate referral indication for fetal echocardiogram (2-4). This will confirm the finding of disproportion, provide indexed values (to gestational age or fetal body size) of cardiac structures (confirming which of the compared structures and at what extend deviate from normative values) (8-11), confirm or detect of associated fetal heart malformations (atrial, ventricular septum defects, abnormalities of inflow and outflow valves). Application of advanced imaging fetal heart imaging can be helpful (12,13). Counselling of the family regarding defect-specific postnatal treatment and long term prognosis will follow (14), as well as recommendation for fetal karyotyping including heart defect specific defects –such as Di George etc, in case where karyotyping has not already been performed.

Summary

Training in the acquisition of the recommended fetal heart views during routine fetal heart imaging is crucial for the early detection of fetal CHD, as most cases do not have an indication for fetal echocardiogram, representing a specialized evaluation reserved for specific indications. Fetal heart chamber and /or great artery disproportion is an easily detected abnormality of the recommended views of the fetal heart during routine mid-trimester ultrasound scan screening. Provided that the finding is not due to fetal unfavorable position, external fetal heart compression or loading conditions, there is an immediate referral indication for fetal echocardiography, as the risk of fetal congenital heart disease is high. ■

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