The Three-Vessels and Tracheal View in Early Prenatal Diagnosis of Cardiac Defects

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Abstract

Objective: To describe our experience with late first and early second trimester, prenatal diagnosis of abnormalities in the three-vessels and tracheal view (3VT) of the heart, utilizing high-resolution ultrasonography and color Doppler techniques.

Materials and Methods: Fetuses with structural heart defects detected at late first and early second-trimester prenatal screening, which were diagnosed with an abnormality on the 3VT. Color Doppler was added in all cases. All patients received genetic counselling and pediatric cardiology consultation.

Results: Twenty-one fetuses had an abnormality detected on the three-vessels and tracheal view. The median gestational age was 15 weeks (range 12-18). Lesions diagnosed included abnormal aortic size (hypoplastic left heart syndrome, narrow aorta, coarctation of the aorta); right aortic arch; abnormal pulmonary artery size (tetralogy of Fallot, pulmonary atresia); abnormal vessels arrangement (transposition of the great arteries); two vessels; four vessels (persistent left superior vena cava); and double outlet right ventricle. Application of color Doppler improved diagnosis in cases involving stenotic or atretic valves and aided in determining ductal flow.

Conclusion: The three-vessels and tracheal view is efficient in late first and early second trimester prenatal diagnosis of cardiac defects, which involve the outflow tracts and the aortic arch, enabling early identification of critical heart defects. Addition of color Doppler aids and further contributes to the diagnosis. We suggest that examination of the three-vessels and tracheal view and color Doppler application should be considered in every low and high-risk prenatal sonographic evaluation.

Keywords: fetal heart, three-vessels and tracheal view, early pregnancy, ultrasound, color Doppler

Özet

Kalp Defektlerinin Erken Prenatal Teşhisinde “Üç Damar ve Trakeal İnceleme”

Amaç: Bu yazıın amacı, yüksek çözünürlüklü ultrasonografi ve renkli Doppler teknikleri kullanarak, geç birinci ve erken ikinci trimesterde kalbin üç damar ve trakeal incelemesiyile (3VT) anomalilerin prenatal teşhisiyle ilgili tecrübelerimizi paylaşmaktır.

Materiyal ve Metot: Birinci ve erken ikinci trimesterde 3VT’de anormal bir görüntü sahtanmasıyla yapsal kalp bozuklukları beliriren fetsüler incelemiştir. Her vakada aynı zamanda renkli Doppler de kullanılmuştur. Tüm hastalara genetik danışmanlık verilmiş ve pediyatrik karniyoloji konsültasyonu yapılmıştır.

Sonuçlar: Fetsülerden 21’inde 3VT’de bir anormallige sahip oldukları gösterdi. Medyan gestasyonel yaş 15 hafta idi (12-18 hafta arası). Teşhis edilen lezyonlar: Anormal aort genişliği (hipoplastik sol kalp sendromu, dar aorta, aortun koarktasyonu), sağ aortik ark, anormal pulmoner arter genişliği (Fallot tetralojisi, pulmoner atrezi), anormal büyük damar dizilimi (büyük
Fetal heart examination is integral to every low and high-risk prenatal sonographic anomaly scan. It is gradually becoming more exact and thorough and accessible to the majority of examiners and patients. Fetal sonographic heart scanning by the four-chamber view alone (1,2) is an excellent screening method, but may miss cardiac anomalies involving the great vessels or outflow tracts (3-8). In order to compensate for the weaknesses of the four-chamber view, other views have been studied to include the ventricular outflow tracts (7,9). The detection rates in the studies cited in the literature vary according to the used approach, the degree of risk of the study population, the expertise of the examiners, and the timing of the examination (8).

The introduction of high-frequency (5-9 MHz) vaginal ultrasound probes has enabled detailed early visualization of cardiac structures, making it possible to diagnose malformations of the fetal heart in the first or early second trimester of pregnancy (10-13). More recent reports utilize specialized high-resolution abdominal and transvaginal transducers, as well as 3D and 4D advanced technologies (14-20). However, these technologies are not widely available to low risk populations in everyday practice, thus necessitating the use of simple specialized 2D views.

The three-vessels view, as suggested by Yoo et al (21,22), is a transverse orthogonal view of the fetal upper mediastinum, which demonstrates the main pulmonary artery (PA), the ascending aorta (Ao), and the superior vena cava (SVC) from left to right in a straight line. Using transvaginal echocardiography the success rate in visualization of the three-vessels view towards the end of the first trimester reaches 97% (23). The three-vessels and tracheal view (7,24,25) adds information on great vessel location in relation to the trachea. The role of the three-vessels and tracheal view in the diagnosis of congenital heart defects has been previously examined in a descriptive study in second and third trimester fetuses (26). On application of color Doppler, the three-vessels and tracheal view proves to be an easier and more reliable means of determining ductal flow direction compared with a long-axis view (26). The three-vessels and tracheal view has been proposed as an adjunct to the classical four chamber view and extended cardiac evaluation, in order to accommodate an effective and accurate examination method into a limited time frame accessible to every patient in busy clinical practices.

To the best of our knowledge, to this date, there has been no study performed evaluating specifically the use of the three-vessels and tracheal view in earlier pregnancy in a mixed low and high-risk population. Therefore, the objective of this study was to describe our experience with prenatal diagnosis of abnormalities in the three-vessels and tracheal view of the fetal heart, diagnosed in the late first or early second trimester, utilizing high-resolution ultrasonography and color Doppler techniques.

Materials and Methods

This study was a retrospective analysis of consecutive cases with an abnormal three-vessels and tracheal view. Our unit services low-risk populations attending routine prenatal anomaly scanning, as well as high-risk patients referred because of suspected fetal anomalies from outpatient community practices, or patients with a known history of congenital heart disease. We screen approximately 2000 patients a year. Fetal echocardiography is included as part of all targeted organ scans. The three-vessels and tracheal view was evaluated during routine ultrasound examination performed to rule out malformations. Gestational age was determined by the beginning of the last menstrual period and verified by sonographic measurement of the crown-rump length in early pregnancy. All women gave written informed consent for screening.

Fetal cardiac examination was standardized using the extended protocol for fetal echocardiography, as previously reported (9,27). The three-vessels and tracheal view was obtained by sliding the transducer upward and slightly oblique from the four-chamber view plane toward the fetal upper mediastinum. In this view, the main pulmonary artery (PA) is seen in direct communication with the ductus arteriosus (DA), to the right of it a transverse section of the aorta (Ao), and a cross section of the superior vena cava (SVC) are arranged in a straight line and in decreasing order in size from left anterior to the right posterior aspect of the mediastinum respectively, the PA being the largest, and the SVC the smallest in diameter (24,25,28). The trachea was subsequently noted in cross section located posteriorly and to the right of the ascending aorta (Figure 1). Color Doppler examination was routinely performed. Examinations were performed transabdominally or transvaginally as necessitated.

All the patients were examined by experienced sonographers (RA, YZ). Ultrasonography was performed using ATL 5000...
nary stenosis. The child is now four years old and has undergone multiple surgeries. In the current pregnancy, she was followed from 12 weeks gestation onwards and was found to have a similar malformation once again. She was offered termination of pregnancy but declined for religious purposes. The offspring was operated after birth and is now 1-year old. The mother and both children tested negative for 22q11 microdeletion and had a normal karyotype. Patient 7 had previously delivered one child with a similar malformation; the child underwent surgery, and is now 7 years old. In a subsequent pregnancy a similar diagnosis was made and she opted for termination of pregnancy. Both child and fetus had a normal karyotype and tested negative for 22q11 microdeletion. She opted for termination of the current pregnancy.

Ten patients had nuchal translucency testing. Four of these (40%) had a nuchal translucency measurement above the 95th percentile for gestational age (patients 3, 5, 17, and 20). An abnormal karyotype was found in three of the four.

Table 2 shows the lesions that were diagnosed by type of three-vessels and tracheal view abnormality. An abnormality in the aorta was seen in eight fetuses (patients 1, 5, 13, 15-19). Four of these had classical hypoplastic left heart syndrome defined as mitral and aortic atresia. One fetus had mitral atresia with double outlet right ventricle and aortic atresia (Fetus 1). Fetus 17 still had forward flow through the aorta and a large ventricular septal defect. Five pregnancies were terminated by parental request. Two of these had an abnormal karyotype, trisomy 18 and 46XO, respectively. Patient 15 was diagnosed with a narrow aorta and a suspected coarctation of the aorta but was consequently lost to follow-up. Patient 19 was diagnosed with a narrow aorta and a persistent left superior vena cava. The baby was delivered, the narrow aorta resolved, and the child is developing normally to date.

An abnormality in the pulmonary artery was observed in seven fetuses (patients 6-8, 10, 11, 14, 20). Five fetuses had tetralogy of Fallot (patients 6, 10, 11, 14, and 20). Four of these pregnancies were terminated by parental request and one continued the pregnancy (patient 20) (Figure 2). Patient 6 had triploidy detected in the karyotype. Patient 8 was diagnosed with pulmonary and tricuspid atresia, and a right aortic arch (Figure 3). The karyotype showed a mosaic with 50% of cells with trisomy 9. The pregnancy was terminated. Patient 7, as described above, had two vessels in the three-vessels and tracheal view, and was diagnosed with transposition of the great arteries with pulmonary atresia and a ventriculo-septal defect.

An abnormal vessel number was observed in seven fetuses. Four vessels were seen in three fetuses with persistent left superior vena cava (patients 9, 10, and 19). The fetus of patient 9 also had an atrioventricular septal defect and hydrops. The patient declined karyotyping for religious reasons, and subsequently delivered a fetus with trisomy 21. Four patients had two vessels in the three-vessels and tracheal view due to severe atresia of a major vessel (patients 1, 6, 7, and 11). The last three requested termination of pregnancy. A double outlet right ventricle was seen in two fetuses (patients 3 and 21). Two patients have a similar malformation once again. She was offered termination of pregnancy but declined for religious purposes. The offspring was operated after birth and is now 1-year old. The mother and both children tested negative for 22q11 microdeletion and had a normal karyotype. Patient 7 had previously delivered one child with a similar malformation; the child underwent surgery, and is now 7 years old. In a subsequent pregnancy a similar diagnosis was made and she opted for termination of pregnancy. Both child and fetus had a normal karyotype and tested negative for 22q11 microdeletion. She opted for termination of the current pregnancy.

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Patient 4 experienced resolution of the findings throughout pregnancy. Patient 12 had numerous additional findings but the situs and pulmonary venous return were normal. She requested pregnancy termination but refused karyotyping. Abnormal position of the aortic arch (Figure 4) was seen in patient 8, who had a right aortic arch (described above). Abnormal vessel arrangement was seen in two fetuses, one with transposition of the great arteries and the other with a right aortic arch.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>GA</th>
<th>CHD</th>
<th>Associated findings</th>
<th>Karyotype</th>
<th>VCF</th>
<th>Follow-up</th>
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<tr>
<td>1</td>
<td>21</td>
<td>12</td>
<td>TGA, AA, MA</td>
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<td>normal</td>
<td>operated, alive</td>
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<tr>
<td>2</td>
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<td>16</td>
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<td>normal</td>
<td>not confirmed</td>
</tr>
<tr>
<td>3</td>
<td>32</td>
<td>14</td>
<td>DORV, VSD, asymmetric ventricles L&gt;R</td>
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<td>Trisomy 18</td>
<td>NP</td>
<td>TOP</td>
</tr>
<tr>
<td>4</td>
<td>31</td>
<td>14</td>
<td>dilated SVC, asymmetric atria</td>
<td>Pelvic kidney, pyelectasis</td>
<td>declined</td>
<td>NP</td>
<td>not confirmed</td>
</tr>
<tr>
<td>5</td>
<td>30</td>
<td>12</td>
<td>AA</td>
<td>increased NT, pyelectasis,</td>
<td>XO</td>
<td>NP</td>
<td>TOP</td>
</tr>
<tr>
<td>6</td>
<td>19</td>
<td>15</td>
<td>TOF</td>
<td>IUGR, DWM, clenched hands, Triploidy</td>
<td>NP</td>
<td>TOP</td>
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</tr>
<tr>
<td>7</td>
<td>36</td>
<td>16</td>
<td>TGA, VSD, PA</td>
<td>none</td>
<td>declined</td>
<td>NP</td>
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<tr>
<td>8</td>
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<td>16</td>
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<td>16</td>
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<td>declined</td>
<td>NP</td>
<td>delivered</td>
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<td>16</td>
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<td>NP</td>
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<td>17</td>
<td>dilated SVC, AVSD, pericardial effusion</td>
<td>cervical cysts, hyperechogenic bowel, clenched fist</td>
<td>NP</td>
<td>NP</td>
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<td>35</td>
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<tr>
<td>14</td>
<td>28</td>
<td>14</td>
<td>TOF</td>
<td>IUGR, cerebellar hypoplasia</td>
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<td>normal</td>
<td>TOP</td>
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<td>18</td>
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<td>27</td>
<td>15</td>
<td>AS, MA, large VSD,</td>
<td>increased NT, large cervical cyst, retrograde flow ductus venosus, clenched hands</td>
<td>trisomy 18</td>
<td>NP</td>
<td>TOP</td>
</tr>
<tr>
<td>18</td>
<td>37</td>
<td>15</td>
<td>AA, MA,</td>
<td>none</td>
<td>normal</td>
<td>normal</td>
<td>TOP</td>
</tr>
<tr>
<td>19</td>
<td>32</td>
<td>14</td>
<td>narrow aorta, left SVC, mildly asymmetric ventricles</td>
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<td>normal</td>
<td>normal</td>
<td>delivered</td>
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<tr>
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<td>28</td>
<td>14</td>
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<td>increased NT</td>
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<td>delivered</td>
</tr>
<tr>
<td>21</td>
<td>33</td>
<td>15</td>
<td>DORV, TGA, VSD, small LV</td>
<td>none</td>
<td>normal</td>
<td>normal</td>
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</tbody>
</table>
the great arteries and one with a double outlet right ventricle (patients 1 and 21). Patient 1 was described above and patient 21 requested pregnancy termination.

Application of color Doppler improved diagnosis directly in 6 patients with stenotic valves (patients 6, 8, and 11 with pulmonary artery abnormality and patients 13, 16, and 18 with aortic abnormality). Color Doppler was helpful in determining ductal flow in an additional 6 patients with atretic valves (patients 1, 5, 7, 10, 14, and 20).

A total of 14 patients opted for pregnancy termination. To the best of our knowledge, there were no missed diagnoses.

**Discussion**

In this study we described our experience with the use of the three-vessels and tracheal view of the fetal heart, and the added value of color flow Doppler in the prenatal diagnosis of congenital heart disease in late first trimester and early second trimester gestation in a mixed low and high risk population. Cardiac anomalies, which involved the great vessels and outflow tracts, were all diagnosed using this method.

As mentioned earlier, in the first trimester low-risk patients, anomalies such as transposition of the great arteries, tetralogy of Fallot, double-outlet right ventricle, and interrupted aortic arch may go undetected if only the four-chamber view is visualized (8). The addition of ventricular outlet views to the four-chamber view increases the detection rate of heart malformations by as much as 30% (9,12). It has been established that most of the lesions involving the ventricular outflow tracts and/or great arteries show an abnormal three-vessels view (21). A recent multi-center study showed a sensitivity of 80% in the detection of congenital malformations by early echocardiography in a high-risk population (29).
The authors described the use of color Doppler, but the three-vessels and tracheal view was not routinely used. In another study, the three-vessels and tracheal view allowed proper recognition of the pulmonary trunk, transverse aortic arch, superior vena cava, and trachea in all but two of 1363 fetuses scanned, requiring an examination time of less than 10 minutes in 98% of the cases (24). The growth of the three cardiac vessels has been evaluated recently and nomograms have been developed for their size during gestation, starting as early as 14 weeks gestation (24,25).

A very recent study (30), described the diagnosis of transposition of the great arteries by sequential segmental transverse views (the four-chamber, five-chamber, three-vessels, and three-vessels and tracheal view) in fetuses throughout pregnancy (13-32 weeks gestation). The authors found that the most reliable clues to the diagnosis were a lateral branching great artery arising from the left ventricle at the level of the five-chamber view, and two vessels rather than three seen at the level of the three-vessels and tracheal view. This study supports the need for wider utilization of the three-vessels and tracheal view.

When analyzing the drawbacks of our study several points come to mind. One is the lack of pathological confirmation for early prenatal diagnoses. The reason for this is that it is our department’s practice to offer pregnancy termination by dilatation and evacuation (D&E) at these gestational weeks. Once offered, most women chose this option without regarding the importance of the pathological diagnosis however heavily stressed. We believe that it is not ethical to preclude offering this procedure for the sole purpose of academic confirmation.

Another issue is the time frame that was chosen for analysis, 12-18 weeks. The reason for this is that, in our country, low risk patients undergo nuchal translucency scanning alone at about 12 weeks and a separate extended fetal scan at 14-16 weeks mostly in outpatient clinical settings. Both of these scans are not government subsidized and are therefore not performed by the entire population. Only one scan at 20-22 weeks is government subsidized. Thus, women chose to have either nuchal translucency or a 14-16 weeks scan or both, based on their pecuniary ability, in addition to the subsidized scan. High-risk patients are usually referred to a tertiary center. For these reasons, some patients have reached our center as late as 18 weeks, after a malformation was suspected in the outpatient setting. Since their referral was a direct result of an earlier scan we decided to include them in our analysis.

Nuchal translucency scanning has enabled early identification of high-risk fetuses at 11-14 weeks of gestation, which are then referred for detailed echocardiography (23,31). In patients with an increased nuchal translucency the finding of a cardiac malformation corroborates with a high frequency of chromosomal aberrations (31,32). In our study, of the ten patients who had a nuchal translucency scan, four were diagnosed with an increased measurement, and three of these were subsequently found to have an abnormal karyotype. Only one of these patients was referred to us directly following the nuchal translucency scan because of septated cystic hygroma. Patient 1 was also diagnosed at 12 weeks but she was a high-risk patient and was therefore followed at our institution from early on. The other patients were either referred later from clinics outside the hospital or initially diagnosed at our center at the 14-16 weeks scan. Of the other patients who were diagnosed before 14 weeks, two had an increased nuchal translucency. Considering the fact that only patients with cystic hygroma or cervical cysts were diagnosed as early as the nuchal translucency scan in clinics outside the hospital, it is not surprising that these fetuses had severe malformations and often an abnormal karyotype.
Twelve patients had some mild or severe extra cardiac findings. The more prominent ones were found to be associated with an abnormal karyotype (five patients). Two patients were diagnosed based on mild extra cardiac findings: mainly pyelectasis. Interestingly in both, findings in the three-vessels and tracheal view were mild and were not confirmed later. There were four patients with mild suspicious findings in the three-vessels and tracheal view (patients 2, 4, 15, and 19). Three were later insignificant or not confirmed, and one patient was lost to follow-up. Three of 21 patients (14%) could thus be considered false positive diagnoses (diagnosed at 14, 16, and 18 weeks gestation) none of which had had an increased nuchal translucency.

In our study, cases, which were diagnosed mainly for the suspicious three-vessels and tracheal view, were usually diagnosed at the 14-16 weeks scan (e.g. patients 6, 7, 10, 11, 14, 20). Only one had an increased nuchal translucency as a cause of referral. This is in accordance with the common practice in our milieu as mentioned previously. Since most of these were low risk patients, it is quite possible that three-vessels and tracheal view aided significantly in reaching the correct diagnosis, which may have otherwise been missed. Low risk patients in outpatient clinics do not always benefit from the advanced 3D and 4D technologies, which are becoming readily available in tertiary referral centers. This supports our belief that the three-vessels and tracheal view should be incorporated into every low risk scan.

A recent systematic review of the accuracy of first-trimester ultrasound examination for detecting major congenital heart disease has found a pooled sensitivity and specificity of 85% (95% CI, 78-90%) and 99% (95% CI, 98-100%), respectively. (33) The authors highlight the strength of the method for diagnosing high-risk mothers in specialized and experienced centers. Allan, in the opinion section of the same issue, states that, “using modern technology, with practice and experience, an accurate diagnosis of congenital heart disease can be made in the first trimester, particularly in the high-risk fetus” (34). Several other authors (23,35) have stated that due to the limitations of early echocardiography it should be reserved for high-risk populations. However, this is not realistic in our milieu, since transvaginal sonographic anomaly scanning is widely performed for low-risk populations and not only in tertiary referral centers. This is precisely why it is imperative that easily obtainable and highly accurate and sensitive diagnostic tools are used. We believe that the three-vessels and tracheal view is just such a diagnostic tool. It provides important clues to the diagnosis of congenital heart defects. Discrepancies in the three-vessels and tracheal view are readily discerned because the visualized anatomy is simple and easily taught.

Color Doppler is an integral part of fetal cardiac evaluation, increasing both the speed and accuracy of the examination (28). It adds hemodynamic information gained throughout the cardiac cycle to morphological imaging. With color Doppler application in the three-vessels and tracheal view,

the convergence of the vessels at the level of the isthmus and ductus arteriosus is characteristically V-shaped, colored blue when the fetal spine is posterior, and red when it is anterior. The addition of color Doppler blood flow in our study further enhanced the diagnostic accuracy in more than half of cases, by aiding in the determination of the direction of flow in the outflow tracts.

In conclusion, a comprehensive fetal echocardiography may detect most significant heart defects, but is considered by many to be time-consuming and demanding special knowledge and skills. It is therefore imperative that accurate, quick, and easily performed scanning views be incorporated into routine examinations, thus improving the diagnostic ability in everyday low-risk clinical practice. We believe that the three-vessels and tracheal view is efficient in early prenatal diagnosis of cardiac defects, which involve the outflow tracts and the aortic arch. It enables early visualization of critical heart defects and early investigation and decision-making. The addition of color Doppler further contributes to the diagnosis. We suggest that visualization of the three vessels and tracheal view and color Doppler application should be considered as an efficient screening tool in every early prenatul sonographic evaluation, in both low and high-risk patients.

References

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