Prenatal Detection of Congenital Heart Defects at the 11- to 13-Week Scan Using a Simple Color Doppler Protocol Including the 4-Chamber and 3-Vessel and Trachea Views

Marcin Wiechec, MD, Anna Knafel, MD, PhD, Agnieszka Nocun, MD

Objectives—The first goal of this study was to analyze the diagnostic performance of the 4-chamber view, 3-vessel and trachea view, and their combination in color mapping during early cardiac evaluations for selecting cases suspicious of congenital heart defects. The second goal was to describe the most common abnormal flow patterns at the levels of the 4-chamber and 3-vessel and trachea views in the late first trimester.

Methods—We conducted a prospective observational study in which a simple cardiac sonographic protocol was applied in fetuses at gestational ages of 11 weeks to 13 weeks 6 days.

Results—A total of 1084 patients with known postnatal or autopsy findings were included in the study. The median maternal age was 32.3 years (range, 27–40 years). The median crown-rump length was 62.2 mm (range, 45–84 mm). Overall, there were 35 cases with a confirmed congenital heart defect (3.22%), including 16 accompanied by aneuploidy. We found that our simple first-trimester cardiac protocol was an effective screening method for congenital heart defects. The most effective approach of the 3 evaluated by us was the combined application of the 4-chamber and 3-vessel and trachea views in color mapping compared to the 4-chamber and 3-vessel and trachea views alone. We defined the most common ventricular inflow patterns and the V sign. The technique we used was simple and easy to reproduce.

Conclusions—We confirmed that evaluation by two basic cardiac views allows for selection of most cases with a univentricular heart, atrioventricular septal defects, coarctation of the aorta, pulmonary stenosis, pulmonary atresia, and conotruncal defects.

Key Words—cardiac defects; fetal echocardiography; fetal heart; first trimester; first-trimester screening; obstetric ultrasound

Received January 31, 2014, from the Chair of Gynecology and Obstetrics, Jagiellonian University, Krakow, Poland. Revision requested March 22, 2014. Revised manuscript accepted for publication July 22, 2014.

Address correspondence to Marcin Wiechec, MD, Chair of Gynecology and Obstetrics, Jagiellonian University, 23 Kopernik St, 31-501 Krakow, Poland.

E-mail: marcin_wiechec@su.krakow.pl

doi:10.7863/ultra.34.4.585

In the past years, plenty of reports about indirect screening methods for the diagnosis of congenital heart defects were published. They were based on markers of aneuploidy, including nuchal translucency, triuspid regurgitation, and ductus venosus velocimetric parameters.[1–4] However, the detection rates for congenital heart defects do not exceed 40% for nuchal translucency above the 95th percentile alone and 58% for nuchal translucency above the same limit combined with a reversed or absent a-wave in the ductus venosus flow profile.[1] Furthermore, indirect methods in

the first trimester are at risk of false-positive findings due to the functional immaturity of the cardiovascular system at this stage of fetal development. It was discovered that transient abnormal fetal cardiac flow patterns could be identified during this period.[5] On the other hand, pulsed wave Doppler venous pulsatility index measurement protocols are difficult to follow by nonexperienced examiners and are at risk of low reproducibility at primary fetal ultrasound centers.[6] Early fetal echocardiography is generally defined as a fetal cardiac scan performed until 16 weeks' gestation.[7,8] It is usually considered a highly specialized scan when the following specific indications are present: increased nuchal translucency measurement, congenital heart defect risk factors, and early diagnosis of extracardiac fetal abnormalities. At our department, however, evaluation of the fetal heart at the time of the nuchal translucency scan has become a routine part of the late first-trimester scan since 2007. According to the literature, this approach increases the detection rate of congenital heart defects in the first trimester to nearly 60% to 80%.[8,9]

Taking into account the performance of indirect screening methods and the low availability of early fetal echocardiography, examiners with less experience expect simple rules describing how to effectively select patients with a high risk of congenital heart defects based not only on the maternal history and secondary signs such as nuchal translucency and triuspid regurgitation but also on direct basic cardiac views. The first goal of this study was to analyze the diagnostic performance of the 4-chamber view, 3-vessel and trachea view, their combination in color mapping, and nuchal translucency during early cardiac evaluations for selecting cases of congenital heart defects. The second goal was to describe the most common abnormal patterns of flow at the levels of the 4-chamber and 3-vessel and trachea views in the late first trimester.

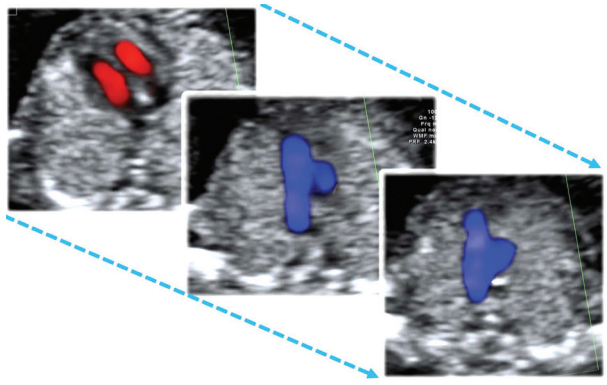
Materials and Methods

The study group included women who underwent sonographic examinations at gestational ages of 11 weeks and 13 weeks 6 days at the ultrasound laboratory of the Chair of Gynecology and Obstetrics, Jagiellonian University, between January 2009 and June 2012. The local Ethics Committee approved the study protocol, and all participating patients gave written consent. This group of **white** pregnant women, who were prospectively examined, went through detailed late first-trimester sonographic screening for aneuploidy, which was performed under condition that the fetal crown-rump length measured between 45 and 84 mm and followed Fetal Medicine Foundation guidelines. The maternal body mass index was calculated in kilograms per square meter on the day of the sonographic examination. The analysis was based on patients who were examined with Voluson E6 ultrasound scanner (GE Healthcare, Zipf, Austria) by experienced physicians (M.W., A.N., and A.K.). The factory presets of the machine were modified to obtain images of high diagnostic quality and to ensure adequate safety limits for an early cardiac scan: the mechanical index for B-mode imaging and the soft tissue thermal index in the color flow mode were set not to exceed a value of 1. All scans were performed with a transabdominal 4–8-MHz hybrid transducer. In difficult scanning conditions, a transvaginal 5–9-MHz hybrid transducer was applied. The above-mentioned standardized settings allowed for accurate cardiac imaging and shortening of the scan duration to fulfill the ALARA (as low as reasonably achievable) principle.[10,11]

The fetal heart examination followed a standardized protocol. In all cases, an operator insonated the fetal chest so that the fetal spine was visualized at the 6- or 12-o'clock position to avoid oblique sections. Then color flow mapping was activated for the shortest time possible. An ideal insonation angle to the interventricular septum of approximately 45° guaranteed easy access to the levels of the 4-chamber and 3-vessel and trachea views, allowing for a smooth transition between these levels (Figure 1).

At the level of the 4-chamber view in color mapping, the following criteria were assessed: number of stripes, subjective impression of their size ratio, and their separation (interventricular septum). At the level of the 3-vessel and trachea view in color mapping, the following criteria were evaluated: number of arterial arms, subjective

Figure 1. Ideal insonation of the beam (45° to the interventricular septum with the fetal spine at the 6-o'clock position) for an early cardiac evaluation starting from the ventricular inflows at the level of 4-chamber view through the 3-vessel view and finishing with depiction of the V sign at the level of the 3-vessel and trachea view.



impression of their size ratio, and flow direction. If only 1 vessel showing an oblique course at the level of the 3-vessel and trachea view was visualized, transposition of great arteries was suspected. In these cases, we extended the study protocol by checking the levels between the 4-chamber and 3-vessel and trachea views in axial sections during transition to search for characteristic patterns of major conotruncal anomalies. Due to the screening purpose of the study, a difficult-to-standardize first-trimester oblique section at the level of the left ventricular outflow tract view, which can demonstrate the parallel course of the great arteries in D-transposition was not included. All patients who continued their pregnancies underwent a complete echocardiographic scan between 18 and 20 weeks' gestation according to American Institute of Ultrasound in Medicine guidelines.[12] In cases with suspicion of congenital heart defects, the final diagnosis was made by postnatal echocardiography or postmortem examinations. Pregnancy outcome data were obtained from medical records. Patients were contacted to provide additional information, if necessary.

Group characteristics were examined by means of descriptive statistics. Subsequently, the sensitivity, specificity, positive predictive value, negative predictive value, and accuracy were calculated separately for each diagnostic algorithm. All calculations were performed with the use of SPSS version 17 data analysis software (IBM Corporation, Armonk, NY).

Results

In total, 1136 patients were examined, but postnatal or autopsy findings were available for only 1084. The median maternal age was 32.3 years (range, 27–40 years); the median body mass index was 22 kg/m^2 (range, 17.5–35 kg/m^2); and the median crown-rump length was 62.2 mm (range, 45–84 mm). The fetal karyotype was determined on the basis of amniocentesis in 168 cases. The remainder of the fetuses (916 cases) were considered euploid based on normal neonatal examination findings. Aneuploidies other than trisomies 21, 18, and 13, monosomy X, and multiple gestations were excluded from the study. The maximum time spent on the cardiac examination did not exceed 5 minutes. Overall, there were 35 cases with a confirmed congenital heart defect (3.22%), including 16 accompanied by aneuploidy (Tables 1 and 2).

In the study population, there were 72 cases of aneuploidy diagnosed, which represented 6.64% of the total population. All scans were performed by a transabdominal approach, and 57 cases (5.25%) required an additional transvaginal evaluation. Nuchal translucency values measured in all cases of congenital heart defects from the study group are shown in Figure 2.

Evaluation by the 4-chamber view alone had sensitivity and specificity of 45.71% and 100%, respectively, compared to 71.43% and 100% for the 3-vessel and trachea view. The combined approach (4-chamber view + 3-vessel and trachea view) yielded the highest sensitivity and specificity of 88.57% and 100% for diagnosis of congenital heart defects at the time of the first-trimester scan (Table 3).

A detailed analysis of the color flow patterns in the 1084 cases examined revealed that at the level of the 4-chamber view, 1068 cases showed 2 ventricular inflows (inflow pattern 1); 4 showed 2 inflows with a dominant inflow that was closer to the anterior thoracic wall (inflow pattern 2); 4 showed initially shared inflows (inflow pattern 3); and 8 showed a singular inflow (inflow pattern 4). Further analysis in the second trimester revealed that in inflow pattern 1, 11 congenital heart defects were identified with a normal karyotype, and an additional 6 were coincident with aneuploidy; in inflow pattern 2, 1 congenital heart defect was found with a normal karyotype and 3 with aneuploidy; in inflow pattern 3, there were no cases with a normal karyotype and 4 cases with trisomy 21; and in inflow

Table 1. Final Cardiac Diagnoses Based on at Least Second-Trimester Echocardiography

Diagnosis	n
Normal	1049
AVSD	6
HLHS	5
D-TGA	4
CoA	3
DORV	3
ToF	3
CAT	2
VSD	2
DILV	1
EA	1
PAIVS	1
PS	1
RAA	1
TA 2A	1
UV	1

AVSD indicates atrioventricular septal defect; CAT, common arterial trunk; CoA, coarctation of the aorta; DILV, double-inlet left ventricle; DORV, double-outlet right ventricle; D-TGA, D-transposition of the great arteries; EA, Ebstein anomaly; HLHS, hypoplastic left heart syndrome; PAIVS, pulmonary atresia with an intact interventricular septum; PS, pulmonary stenosis; RAA, right aortic arch; TA 2A, tricuspid atresia with transposed great arteries and pulmonary atresia; ToF, tetralogy of Fallot; UV, univentricular heart; and VSD, ventricular septal defect.

Table 2. Cardiac Sonographic Findings in the First and Second Trimesters With Postnatal Echocardiographic or Autopsy Confirmation in Cases With Congenital Heart Defects

Case	Gestational Age, wk: Sonographic Findings		Confirmation
	1st Trimester	2nd Trimester	
1	12: abnormal 3VTV, 3VV	18: D-TGA	Postnatal
2	13: abnormal 3VTV, 3VV	19: D-TGA	Postnatal
3	12: abnormal 3VTV, 3VV	19: D-TGA	Postnatal
4	12: abnormal 3VTV, 3VV	20: D-TGA	Postnatal
5	13: abnormal 4CV, 3VTV, 3VV	20: ToF	Postnatal
6	12: abnormal 3VTV	19: ToF	Postnatal
7	12: abnormal 3VTV	18: ToF	Postnatal
8	13: abnormal 3VTV	19: DORV	Autopsy
9	12: abnormal 3VTV	21: DORV	Autopsy
10	11: abnormal 3VTV	18: DORV	Autopsy
11	11: normal	20: AVSD	Postnatal
12	12: normal	19: AVSD	Postnatal
13	12: abnormal 4CV, 3VTV	20: CAT	Postnatal
14	12: abnormal 3VTV	21: CAT	Postnatal
15	12: abnormal 3VTV	20: PAIVS	Postnatal
16	13: abnormal 3VTV	21: PS	Postnatal
17	12: abnormal 3VTV	18: RAA	Postnatal
18	13: normal	21: VSD	Postnatal
19	12: normal	20: VSD	Postnatal
20	13: abnormal 4CV, 3VTV	19: CoA	Autopsy
21	12: abnormal 4CV, 3VTV	20: CoA	Autopsy
22	13: abnormal 4CV, 3VTV	19: CoA	Autopsy
23	11: abnormal 4CV	19: EA	Autopsy
24	12: abnormal 4CV	20: AVSD	Postnatal
25	12: abnormal 4CV	19: AVSD	Postnatal
26	13: abnormal 4CV	20: AVSD	Postnatal
27	12: abnormal 4CV	20: AVSD	Postnatal
28	12: abnormal 4CV, 3VTV	19: HLHS	Postnatal
29	11: abnormal 4CV, 3VTV	19: HLHS	Postnatal
30	12: abnormal 4CV, 3VTV	20: HLHS	Postnatal
31	12: abnormal 4CV + 3VTV	21: HLHS	Postnatal
32	13: abnormal 4CV + 3VTV	18: HLHS	Postnatal
33	12: abnormal 4CV + 3VTV	20: TA 2A	Autopsy
34	12: abnormal 4CV + 3VTV	21: DILV	Postnatal
35	13: abnormal 4CV	20: UV	Autopsy

4CV indicates 4-chamber view; 3VTV, 3-vessel and trachea view; and 3VV, 3-vessel view; other abbreviations are as in Table 1.

pattern 4, 6 congenital heart defects were identified with a normal karyotype and 2 with a chromosomal aberration. The details are presented in Table 4.

At the level of the 3-vessel and trachea view in color mapping, 1059 cases showed equal arms of the V sign (type 1); 3 showed a lower-intensity arm that corresponded to a transverse section through the aortic arch (type 2); 2 showed a lower-intensity arm that corresponded to a transverse section through the ductal arch (type 3); 5 showed a singular arm with a vertical course (type 4); 14 showed a singular arm with an oblique course (type 5); and 1 showed a U-shaped V sign (type 6). Details are depicted in Table 5.

A detailed analysis in the second trimester allowed for diagnosis of congenital heart defects in particular types of first-trimester V signs. The findings included 2 cases of congenital heart defects with a normal karyotype and an additional 8 with confirmed aneuploidy in type 1, 3 cases with Turner syndrome in type 2, 2 cases with a normal karyotype in type 3, 4 cases with euploidy and 1 with trisomy 18 in type 4, 10 cases with a normal karyotype and 4 with chromosomal defects in type 5, and 1 case with a normal karyotype in type 6.

Figure 2. Nuchal translucency measurements in cases with congenital heart defects from the study population. CRL indicates crown-rump length; and NT, nuchal translucency.

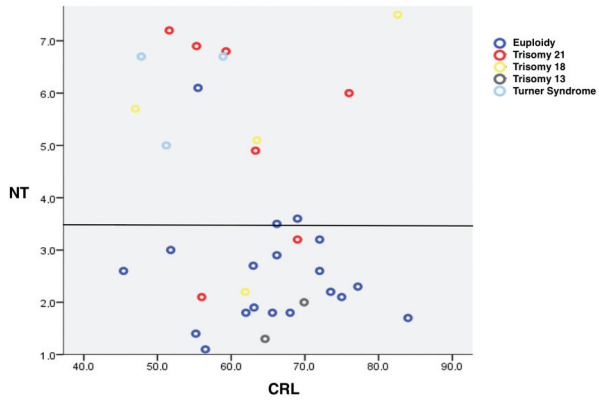
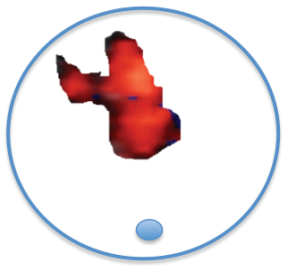


Table 3. Screening Performance of Each Method for Congenital Heart Defects

Parameter	4CV	3VTV	4CV + 3VTV	NT >3.5 mm	NT >95th Percentile
Sensitivity, %	45.71 (30.47–61.81)	71.43 (54.94–83.67)	88.57 (74.05–95.46)	37.14 (23.17–53.66)	60.00 (43.57–74.45)
Specificity, %	100 (99.64–100)	100 (99.64–100)	100 (99.64–100)	95.61 (94.2–96.7)	89.51 (87.51–91.23)
PPV, %	100 (80.64–100)	100 (86.68–100)	100 (88.97–100)	22.03 (13.35–34.13)	16.03 (10.73–23.27)
NPV, %	98.22 (97.24–98.86)	99.06 (98.27–99.49)	99.62 (99.03–99.85)	93.73 (92.12–95.02)	98.53 (97.55–90.32)
Accuracy, %	98.25 (97.28–98.88)	99.08 (98.31–99.5)	99.63 (99.06–99.86)	93.73 (92.12–95.02)	88.56 (86.53–90.32)

Values in parentheses are 95% confidence intervals. NPV indicates negative predictive value; NT, nuchal translucency; and PPV, positive predictive value; other abbreviations are as in Table 2.

Table 4. Inflow Patterns in Diastole at the Level of the 4-Chamber View and the Corresponding Heart Defects in the Studied Population

Inflow Pattern 1 (n = 1068)	Inflow Pattern 2 (n = 4)	Inflow Pattern 3 (n = 4)	Inflow Pattern 4 (n = 8)
			

Defect	n	Defect	n	Defect	n	Defect	n
D-TGA	4	CoA	3	AVSD	4	HLHS	5
DORV	3	EA	1			TA 2A	1
ToF	3					DILV	1
AVSD	2					UV	1
CAT	2						
VSD	2						
PAIVS	1						
PS	1						
RAA	1						
None	1049						

Abbreviations are as in Table 1.

Table 5. Breakdown of 1084 Fetuses Scanned Between 11 Weeks and 13 Weeks 6 Days According to V-Sign Type at the Level of the 3-Vessel and Trachea View on Color Mapping

3VTV 1 (n = 1059)	3VTV 2 (n = 3)	3VTV 3 (n = 2)	3VTV 4 (n = 5)	3VTV 5 (n = 14)	3VTV 6 (n = 1)
					

Defect	n	Defect	n	Defect	n	Defect	n	Defect	n	Defect	n
AVSD	6	CoA	3	PS	1	HLHS	5	D-TGA	4	RAA	1
VSD	2			PAIVS	1			DORV	3		
UV	1							ToF	3		
EA	1							CAT	2		
None	1049							DILV	1		
								TA 2A	1		

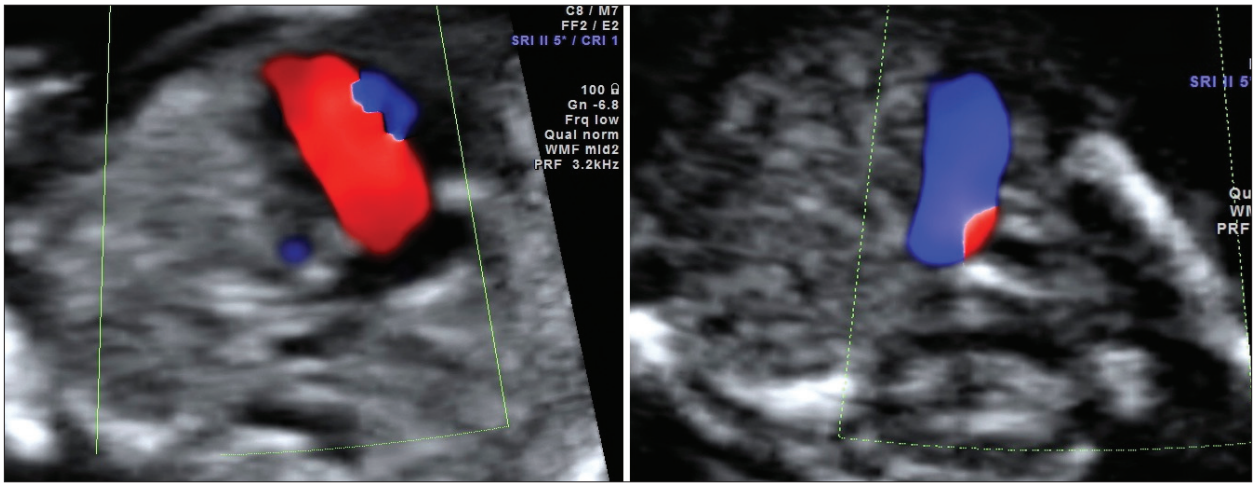
Abbreviations are as in Table 1.

In this study, we tried to analyze the most common patterns on color mapping that can be encountered in congenital heart defects. Figures 3–9 depict first-trimester patterns of congenital heart defects found in our study population.

Discussion

There have been many studies published to date that focused on detection of congenital heart defects at the time of the nuchal scan based on sonographic markers such as nuchal translucency, triuspid regurgitation, and the ductus venosus.[1,4,13,14] To our knowledge, this article is the first practical description of a straightforward and direct screening method for congenital heart defects in the first trimester. In this study we found that our simple first-trimester cardiac protocol was an effective screening method for congenital heart defects. The most effective approach of the 3 direct methods evaluated by us was the combined application of the 4-chamber and 3-vessel and trachea views in color mapping compared to the 4-chamber and 3-vessel and trachea views alone. The combined approach had sensitivity of 88.57% and specificity of 100% for detection of congenital heart defects in the unselected population studied. These results are consistent with the observations of Iliescu et al,[15] who examined 5472 unselected cases between gestational ages of 12 weeks and 13 weeks 6 days by using a color cardiac sweep and showed a 90% detection rate for major congenital heart defects. In that study, 67.1% of cases with major anomalies did not reveal increased nuchal translucency.[15] Similarly, Perisco et al[16] showed a 93.1% detection rate for congenital heart defects in a group of 886 patients including 100 cardiac anomalies. Also, Volpe et al[17] demonstrated that the direct assessment of the heart at 11 to 14 weeks is a feasible method of screening for congenital heart defects in an unselected population. Direct assessment of the fetal heart in the first trimester to search for congenital heart defects in a high-risk population revealed that the examiner's experience and the selection of patients are among the most important factors that have an impact on detection of congenital heart defects.[16,18,19] The two cardiac views we used can be easily adopted in a nuchal scan, as, according to our observations, the most common fetal position in the late first trimester is the transverse one, which increases the chance of successful visualization of basic cardiac views on color mapping within a few minutes. A simple direct method of cardiac evaluation is superior to the simplest first-trimester screening tool for congenital heart defects of using nuchal translucency thickness. Westin et al[2] reported an extremely low detection rate for nuchal translucency of greater than 3.5 mm for congenital heart defects at the level of 5.8% based on a low-risk population of 16,383 patients. In our study population, a higher figure of 37.14% was obtained due to the high number of chromosomal aberrations observed and the analysis of a mixed-risk population.

Figure 3. A case of hypoplastic left heart syndrome at 12 weeks. The 4-chamber view (left) shows a color stripe on the right side, which represents filling of the right ventricle. In the 3-vessel and trachea view (right), a broad arterial arm with a course parallel to the ultrasound beam (vertical course) is visible and represents a dilated pulmonary artery and ductus arteriosus. Although the 2 arms of the V sign can be seen, their sizes are severely unequal, and the flow within the aortic arch is abnormal, showing retrograde flow on the lower right side (retrograde filling toward a hypoplastic aortic arch from the ductus arteriosus), which is usually difficult to visualize on B-mode imaging without color mapping.



The strength of our study was the fact that the evaluation of the fetal heart was performed in a high number of patients who underwent screening sonography with known follow-up findings. Combined assessment with the 4-chamber and 3-vessel and trachea views in color mapping allowed the diagnosis of various types of cardiac anomalies. Furthermore, this method was applied by 3 independent examiners. A possible weakness of the study might be the fact that all of the examiners had extensive expertise in cardiac evaluation and early anomaly assessment using high-end ultrasound equipment. Second, because of the screening nature of the study, the total number of congenital heart defects was not very high.

To our knowledge, there is a lack of reports in the literature that compared the efficacy of congenital heart defect screening with the 4-chamber view, the 3-vessel and trachea view, and the combined method in the first trimester. Studies evaluating an extended cardiac examination, including the 4-chamber view and outflow tracts, in the second trimester in screening for congenital heart defects have shown its better performance compared to the 4-chamber view alone.[20]

Additionally, in our study, we defined the most typical patterns of congenital heart defects on color Doppler imaging at the level 4-chamber and 3-vessel and trachea views. Information collected from a simple cardiac protocol based on the evaluation of ventricular inflow patterns enabled identification of a univentricular heart (all cases), coarctation of the aorta in Turner syndrome (all cases), and most atrioventricular septal defects (4 of 6). Assessment of the V sign allowed for selection of cases with coarctation of the aorta in Turner syndrome, cases of pulmonary stenosis, pulmonary atresia with an intact intraventricular septum, hypoplastic left heart syndrome, a right aortic arch, conotruncal abnormalities, and transposed great arteries in a double-inlet left ventricle and tricuspid atresia type

Figure 4. D-Transposition of the great arteries at 12 weeks: 4-chamber view (4CV), 3-vessel view (3VV), and 3-vessel and trachea view (3VTV) of D-transposition of the great arteries (1–3) and a normal heart (4–6). In isolated D-transposition of the great arteries, the 4-chamber view (1) typically shows normal-appearing anatomy, with 2 symmetric ventricles, a continuous ventricular septum, and insertion of the atrioventricular valves. On color mapping, 2 stripes are visualized, which represent normal filling of the cardiac chambers during diastole. In the 3-vessel and trachea view (3), color Doppler imaging shows only 1 vessel in an oblique fashion instead of 2. The image of a single vessel that takes a wide sweep on leaving the right ventricle should always raise the suspicion of transposition of the great arteries. The pulmonary artery is not visible in this plane due to the inferior and posterior relationships with the aorta in this condition. This factor is why the next step should be a subtle tilt backward toward the 4-chamber view to the level of the classic 3-vessel view (2). In this view, 2 separate arterial signals are shown, and 1 of these vessels (right, representing the aorta) is located more anteriorly.

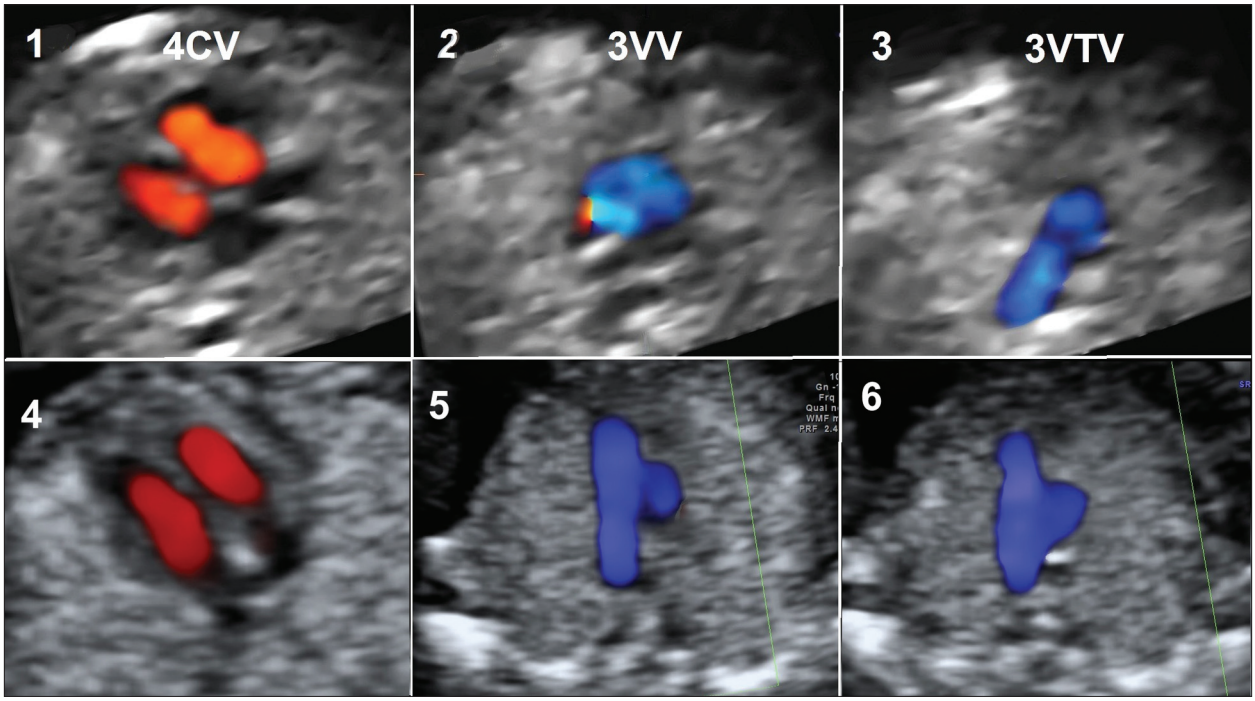


Figure 5. Case that was confirmed at midgestation as a double-inlet left ventricle with transposed great arteries at 13 weeks. At the level of the 4-chamber view, a singular inflow was found; the 3-vessel and trachea view shows a singular arterial vessel in an oblique fashion; an approximate level of the right ventricular outflow tract (RVOT) between the 4-chamber and 3-vessel and trachea views showed 2 arterial signals, with a more anterior position of the right-sided signal. The 3-vessel and trachea view is similar to that of simple D-transposition of the great arteries. Abbreviations are as in Figure 4.

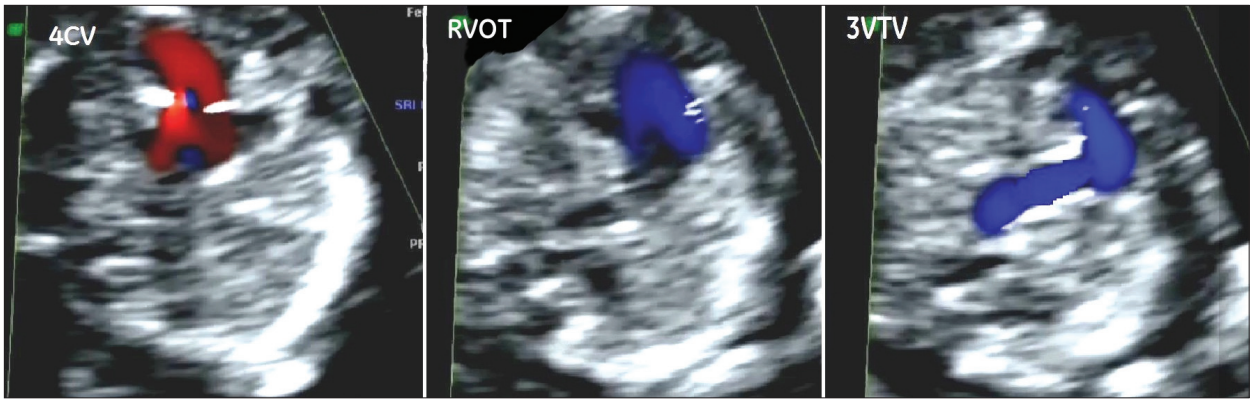


Figure 6. Complex congenital heart defects confirmed at midgestation as tricuspid atresia type 2A at 12 weeks' gestation. Suspicious findings in the first trimester were as follows: a singular inflow at the level of the 4-chamber view, a singular arterial vessel with a slightly oblique course at the level of the 3-vessel and trachea view, and 2 separate arterial color Doppler signals at the level of the 3-vessel view, arranged in a way that the right-sided vessel was more anterior, and retrograde flow was seen in the left-sided vessel. Abbreviations are as in Figure 4.

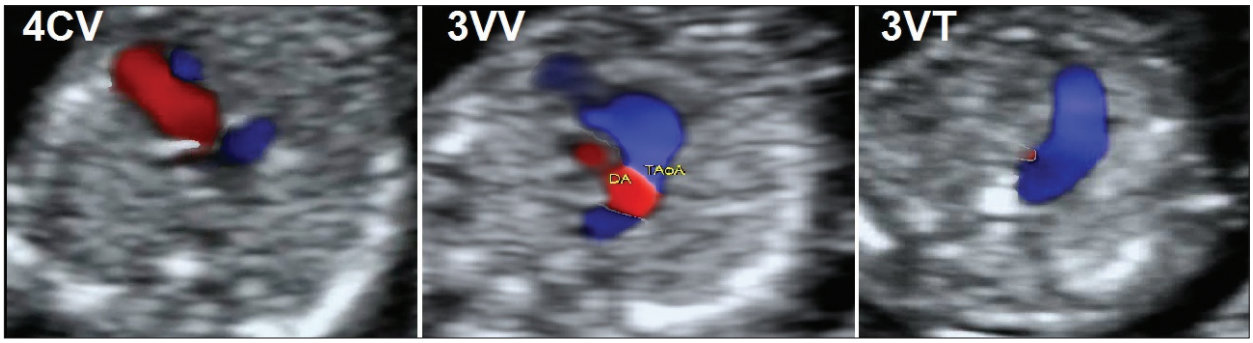
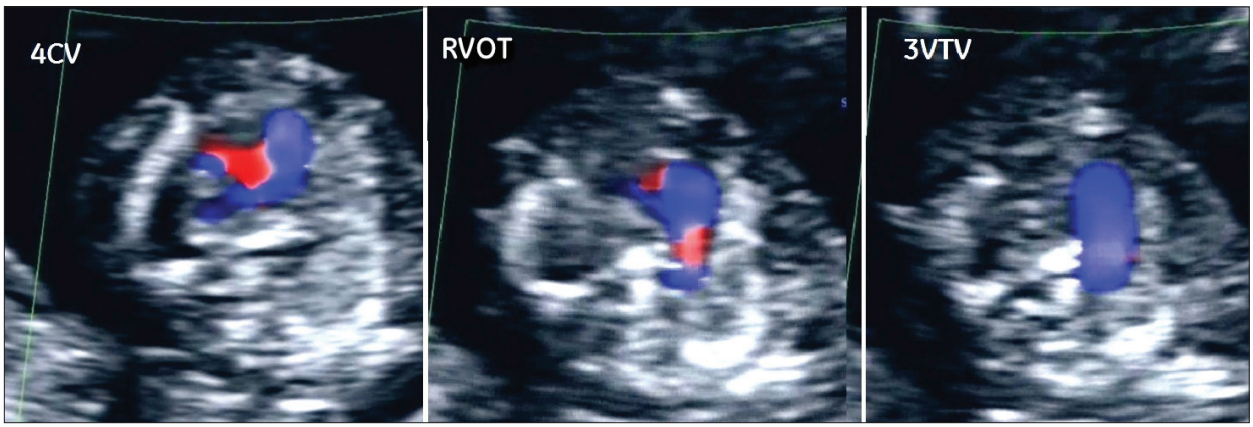


Figure 7. Cardiac anatomy suspicious for a conotruncal defect in the late first trimester (12 weeks). Deviation of the cardiac axis is shown at the level of the 4-chamber view; a singular broad arterial vessel is shown at the level of the 3-vessel and trachea view; and 2 arterial signals with unbalanced intensity (right-sided signal is stronger) is shown on the right ventricular outflow tract view. This case was confirmed in the second trimester as tetralogy of Fallot with a right-sided aortic arch. Abbreviations are as in Figures 4 and 5.



2A. The 3-vessel and trachea view was found to be very sensitive for conotruncal abnormalities when only 1 oblique arm of the V sign was observed. However, it cannot be used for a conclusive differential diagnosis. In such cases, we suggest taking particular care to check the axial levels between the 4-chamber and 3-vessel and trachea views.

Our simple approach to basic cardiac views in the first trimester is similar to the second-trimester observations described by Chaoui and McEwing.[21] They described color Doppler findings for various congenital heart defects based on the flow patterns seen at the levels of the 4-chamber view, 5-chamber view, and 3-vessel view. Due to the better resolution in the second trimester and analyses performed in both systole and diastole, more information can be obtained later in gestation, but key simple abnormalities show the same typical color mapping images in the first and second trimesters of pregnancy, including atrioventricular septal defects, nonevolutional forms of hypoplastic left heart syndrome, D-transposition of the great arteries, tetralogy of Fallot, a common arterial trunk, tricuspid atresia, a double-inlet left ventricle, and severe forms of Ebstein anomaly, coarctation of the aorta, and pulmonary stenosis.

Figure 8. Cardiac anatomy suspicious for a conotruncal defect in the late first trimester (13 weeks). Deviation of the cardiac axis is shown at the level of the 4-chamber view; 1 large overriding vessel with turbulent flow is shown at the level of the 5-chamber view (5CV); and a singular arterial vessel with very strong convexity, with an almost horizontal course, is shown at the level of the 3-vessel and trachea view. This case was confirmed in the second trimester as a common arterial trunk. Abbreviations are as in Figure 4.

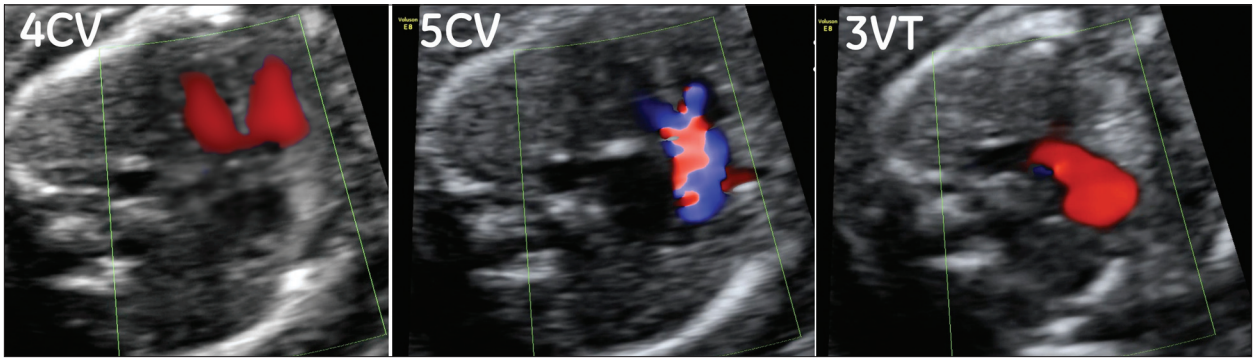
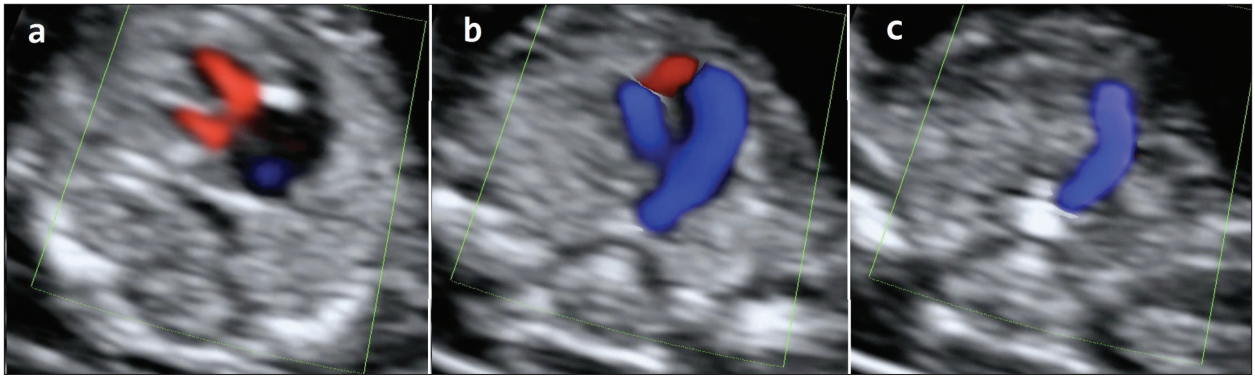


Figure 9. Double-outlet right ventricle at 12 weeks. At the level of the 4-chamber view (**a**), right heart dominance is shown, together with the communication between the ventricles. At the level between the 4-chamber and 3-vessel and trachea views (**b**), a Y-shaped communication between the arches is shown, with the weaker intensity of the signal representing the main pulmonary artery and the arterial duct (left-sided arm). At the level of 3-vessel and trachea view (**c**), a singular arm of the V sign is shown.



References

1. Khalil A, Nicolaides KH. Fetal heart defects: potential and pitfalls of first-trimester detection. *Semin Fetal Neonatal Med* 2013; 18:251–260.
2. Westin M, Saltvedt S, Bergman G, Almström H, Grunewald C, Valentin L. Is measurement of nuchal translucency thickness a useful screening tool for heart defects? A study of 16,383 fetuses. *Ultrasound Obstet Gynecol* 2006; 27:632–639.
3. Maiz N, Plasencia W, Dagklis, T, Faros E, Nicolaides K. Ductus venosus Doppler in fetuses with cardiac defects and increased nuchal translucency thickness. *Ultrasound Obstet Gynecol* 2008; 31:256–260.
4. Pereira S, Ganapathy R, Synegelaki A, Maiz N, Nicolaides KH. Contribution of fetal tricuspid regurgitation in first-trimester screening for major cardiac defects. *Obstet Gynecol* 2011; 117:1384–1391.
5. Bronshtein M, Zimmer EZ, Blazer S, Blumenfeld Z. Transient abnormal fetal cardiac flow patterns at 13 to 17 gestational weeks. *Prenat Diagn* 2012; 32:444–449.
6. Carvalho JS. Screening for heart defects in the first trimester of pregnancy: food for thought. *Ultrasound Obstet Gynecol* 2010; 36:658–660.
7. Yagel S, Weissman A, Rotstein Z, et al. Congenital heart defects: natural course and in utero development. *Circulation* 1997; 96:550–555.
8. Smrcek JM, Berg C, Geipel A, et al. Detection rate of early fetal echocardiography and in utero development of congenital heart defects. *J Ultrasound Med* 2006; 25:187–196.
9. Becker R, Wagner RD. Detailed screening for fetal anomalies and cardiac defects at the 11–13-week scan. *Ultrasound Obstet Gynecol* 2006; 27:613–618.
10. Toms DA. The mechanical index, ultrasound practices, and the ALARA principle. *J Ultrasound Med* 2006; 25:560–561.
11. Salvesen K, Lees C, Abramowicz JS, Brezinka C, Ter Haar G, Maršál K; Board of International Society of Ultrasound in Obstetrics and Gynecology (ISUOG). ISUOG statement on the safe use of Doppler in the 11 to 13 + 6-week fetal ultrasound examination. *Ultrasound Obstet Gynecol* 2011; 37:628.
12. American Institute of Ultrasound in Medicine. AIUM practice guideline for the performance of fetal echocardiography. *J Ultrasound Med* 2013; 32:1067–1082.
13. Sotiriadis A, Papatheodorou S, Eleftheriades M, Makrydimas G. Nuchal translucency and major congenital heart defects in fetuses with normal karyotype: a meta-analysis. *Ultrasound Obstet Gynecol* 2013; 42:383–389.
14. Martinez JM, Comas M, Borrell A, et al. Abnormal first-trimester ductus venosus blood flow: a marker of cardiac defects in fetuses with normal karyotype and nuchal translucency. *Ultrasound Obstet Gynecol* 2010; 35:267–272.
15. Iliescu D, Tudorache S, Comanescu A, et al. Improved detection rate of structural abnormalities in the first trimester using an extended examination protocol. *Ultrasound Obstet Gynecol* 2013; 42:300–309.
16. Perisco N, Moratalla J, Lombardi CM, Zidere V, Allan L, Nicolaides KH. Fetal echocardiography at 11–13 weeks by transabdominal high-frequency ultrasound. *Ultrasound Obstet Gynecol* 2011; 37:296–301.
17. Volpe P, Ubaldo P, Volpe N, et al. Fetal cardiac evaluation at 11–14 weeks by experienced obstetricians in a low-risk population. *Prenat Diagn* 2011; 31:1054–1061.
18. Lombardi CM, Bellotti M, Fesslova V, Cappellini A. Fetal echocardiography at the time of the nuchal translucency scan. *Ultrasound Obstet Gynecol* 2007; 29:249–257.
19. Bellotti M, Fesslova V, De Gasperi C, et al. Reliability of the first-trimester cardiac scan by ultrasound-trained obstetricians with high-frequency transabdominal probes in fetuses with increased nuchal translucency. *Ultrasound Obstet Gynecol* 2010; 36:272–278.
20. Ogge G, Gagliotti P, Maccanti S, Faggiano F, Todros T. Prenatal screening for congenital heart disease with four-chamber and outflow-tract views: a multicenter study. *Ultrasound Obstet Gynecol* 2006; 28:779–784.
21. Chaoui R, McEwing R. Three cross-sectional planes for fetal color Doppler echocardiography. Ultrasound Obstet Gynecol 2003; 21:81–93.