Three cross-sectional planes for fetal color Doppler echocardiography

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ABSTRACT

Routine use of color Doppler during every fetal cardiac examination remains controversial. Many examiners still believe that color should be reserved for cases of suspected congenital heart defect (CHD). In our opinion, color Doppler should be applied in every cardiac scan due to the increase in speed and accuracy that it allows. The purpose of this review is to first explain how color Doppler presets can be optimized and, second, to propose the use of three cross-sectional planes to simplify color Doppler fetal echocardiography: the four-chamber (4CV), five-chamber (5CV) and three-vessel (3VV) views. A practical approach to the detection of CHD with these planes is presented, with typical findings and possible abnormalities evident during systole and diastole. The diastolic pattern on the 4CV is characterized by two equal color stripes. Connection ('H'-sign) or size inequality of the two stripes, or a unilateral color stripe, are important abnormal findings. In systole valve regurgitation should be excluded. In the 5CV, turbulent flow, ventricular septal defect or an overriding aorta ('Y'-sign) can be detected. In the 3VV the aorta and pulmonary trunk should be of nearly equal size and demonstrate antegrade flow. Abnormal findings encountered include absence of one vessel, discrepant size of the vessels, retrograde flow in one of the vessels, or the 'U'-sign, where the trachea is enclosed between both vessels, suggesting right-sided aortic arch. In summary, we propose that color Doppler examination utilizing these three planes alone is sufficient to obtain adequate information for the detection of most common CHD. Copyright © 2002 ISUOG. Published by John Wiley & Sons, Ltd.

INTRODUCTION

Color Doppler echocardiography was introduced into clinical practice in the mid-1980s and was directly applied to fetal echocardiography. Few papers have been published on the efficacy of color Doppler in fetal cardiac defects but clinical experience demonstrates that the technique completes the real-time assessment of heart defects[1–10]. Copel *et al.*[6] found in 1991 that the addition of color flow mapping was essential for accurate diagnosis in 29% of cases, particularly where there was outflow obstruction of the great vessels, and helpful, but not essential, in a further 47% of cases. A decade later, although all current mid- and high-resolution ultrasound machines offer color Doppler, many examiners reserve its use for cases where a cardiac defect is suspected on initial real-time examination. Centers specializing in fetal echocardiography, however, treat color Doppler as an integral component of fetal cardiac evaluation, and are aware that it increases both the speed and accuracy of the examination. The aim of this review is to present a simplified approach to the color Doppler examination of the fetal heart by using three cross-sectional planes. The normal findings and typical abnormalities that can be demonstrated in these planes are presented.

In this review the abnormal findings are presented according to the color Doppler appearances in the three views. The findings in specific heart defects have been described elsewhere[11] and are summarized in Table 1.

COLOR DOPPLER PRESETS FOR FETAL ECHOCARDIOGRAPHY

Optimization of both real-time examination and of color Doppler presets are essential prerequisites for fetal cardiac assessment. Examiners should familiarize themselves with the standard features available on every machine prior to the application of color Doppler as described below.

Velocity scale (or pulse repetition frequency) (Figure 1) allows the examiner to determine the range of velocities in the region of interest. For structures where high velocity flow is expected, such as the atrioventricular (AV) and

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REVIEW

Table 1 Color Doppler findings in common cardiac anomalies

Cardiac defect	Four-chamber view	Five-chamber view	Three-vessel view
Tricuspid atresia with VSD Absent connection between RA and RV; hypoplastic RV, VSD present	Absent flow from RA to RV; blood from RA flows across foramen ovale to LA, and then in diastole to LV; unilateral perfusion across LV inflow tract; left to right shunt across the VSD into small RV	Non-contributory or depending on the ventriculo-arterial connections	Pulmonary stenosis often evident with antegrade flow
Tricuspid dysplasia and Ebstein's anomaly TV dysplasia: thickened valve leaflets Ebstein's anomaly: apical displacement of TV leaflets within the RV RA dilatation common; gross cardiomegaly in severe cases	Severe TR Spectral Doppler can be used to measure pressure gradient and duration of regurgitation	Non-contributory	Tiny pulmonary artery when associated obstruction of RV outflow tract: antegrade flow in pulmonary stenosis; retrograde flow in severe forms
Pulmonary atresia with intact ventricular septum RV hypoplastic, normal-sized or, rarely, dilated; poor contractility of RV; reduced TV movement; atretic pulmonary valve	Reduced or absent tricuspid flow; TR may be evident during systole; rule out ventriculo-coronary fistula	Non-contributory	Lack of antegrade flow across pulmonary valve; retrograde flow through DA; pulmonary trunk smaller caliber than ascending aorta
Pulmonary stenosis Narrowing of semilunar valves; poststenotic dilatation of pulmonary trunk; hypokinetic and hypertrophied RV in severe cases	Tricuspid insufficiency in severe cases or often in third trimester	Non-contributory	Antegrade turbulent flow in pulmonary trunk; rarely retrograde flow through DA in severe cases
Aortic stenosis Narrowing of semilunar valves; poststenotic dilatation of the ascending aorta	Non-contributory	Antegrade turbulent flow with aliasing is characteristic; pulsed Doppler shows high velocities (> 2 m/s)	Turbulent flow in dilated proximal aortic arch
Hypoplastic left heart syndrome/critical aortic stenosis with LV dysfunction Aortic valve atretic or severely stenotic; LV small, normal sized or dilated, but non-contractile; mitral valve atretic or stenotic	Unilateral perfusion of RV; reduced or absent diastolic filling of LV; abnormal left to right shunt across interatrial septum	Hypoplastic aorta, often with retrograde flow; in critical stenosis, antegrade turbulent flow may be observed	Retrograde perfusion in hypoplastic aortic arch
Coarctation of the aorta Narrowing of distal aortic arch or of the whole arch in tubular hypoplasia; LV may be smaller than RV; VSD may be present in some cases with tubular hypoplasia	LV may be smaller than RV	Antegrade flow across the aortic valve; aorta may be of small caliber	Aortic arch smaller than ductal arch; aortic arch flow usually antegrade, unless severe
Ventricular septal defect Defect usually involves perimembranous septum, occasionally inlet or muscular septum; defects larger than 3 mm can be identified with two-dimensional ultrasound	Color Doppler identifies small muscular VSDs and confirms larger VSD	Color flow across perimembranous defect	Non-contributory

Table 1 (*Continued*)

Cardiac defect	Four-chamber view	Five-chamber view	Three-vessel view
Atrioventricular septal defect Septal valve leaflets deformed or absent, with common AV junction and deficient crux	'H'-shaped biventricular diastolic flow across left and right inflow tracts with communication at level of AV valves; TR and MR during systole	Non-contributory	Non-contributory
Tetralogy of Fallot VSD; overriding aorta; infundibular pulmonary stenosis; RV hypertrophy (not evident prenatally)	Non-contributory in most cases	'Y'-shaped systolic flow from both ventricles into overriding aorta	Antegrade (turbulent) flow in tiny pulmonary trunk; in severe cases retrograde flow through DA
Double-outlet RV Aorta and pulmonary trunk arise from RV; LV may be smaller than RV; VSD	VSD with left-to-right shunt may be present; small LV may be evident	Color flow within aorta and pulmonary trunk arising from RV, usually with a parallel course	Obstruction of aortic or pulmonary outflow tracts may be seen
Complete transposition of the great arteries Aorta arises from RV and pulmonary trunk from LV	Non-contributory	Parallel course of great vessels; subpulmonary VSD may be detected	Usually one great vessel seen (aorta)
Truncus arteriosus communis Single great artery, often overriding the interventricular septum, bifurcating into aorta and pulmonary trunk in Type I	Non-contributory	Common arterial trunk seen as a single large vessel overriding interventricular septum; truncus insufficiency may be present	DA often absent; tiny pulmonary artery

AV, atrioventricular; DA, ductus arteriosus; LA, left atrium; LV, left ventricle; MR, mitral regurgitation; RA, right atrium; RV, right ventricle; TR, tricuspid valve regurgitation; TV, tricuspid valve; VSD, ventricular septal defect.
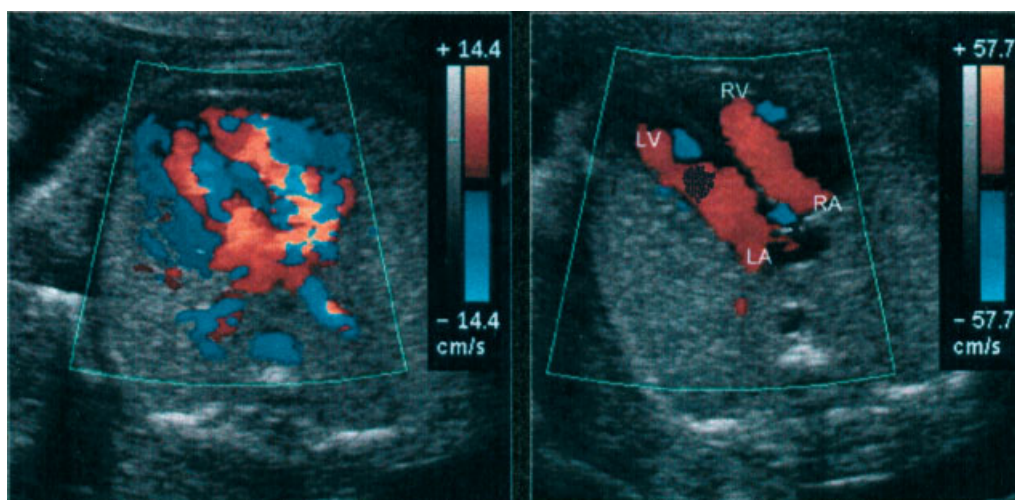
Figure 1 Velocity scale. The same four-chamber view is demonstrated with different velocity scales (pulse repetition frequencies). The left image was obtained using a velocity of ± 15 cm/s, too low for atrioventricular valve perfusion. Color imaging shows aliasing. The image on the right utilized a velocity of ± 57 cm/s, an appropriate range, showing homogeneous perfusion (RA, LA, right and left atrium; RV, LV, right and left ventricle).

semilunar valves and great vessels, a high velocity scale ranging from 40 to 70 cm/s should be selected. The choice of low velocity will demonstrate aliased flow (Figure 1 left) and lead to misinterpretation of findings. Smaller vessels, such as the pulmonary and caval veins, exhibit velocities of about 20 cm/s, suitable for mid-range velocity scale.
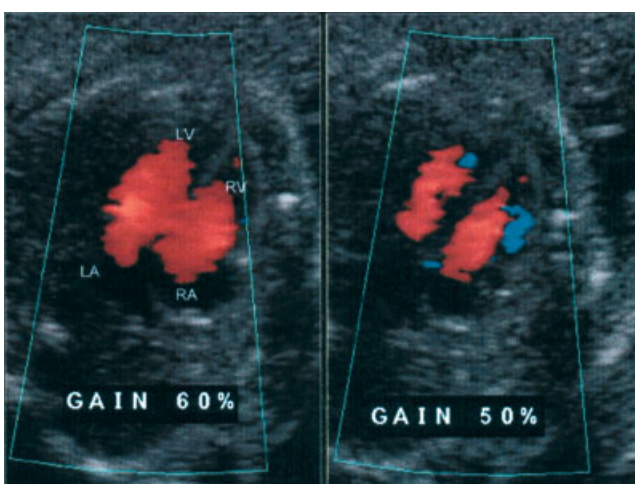
Filter allows elimination of signal from wall movements and low velocities. A high filter is appropriate for cardiac assessment. Visualization of small, low velocity vessels, e.g. pulmonary arteries and veins, is facilitated by selection of a low filter.

Color persistence allows information from previous images to be overlapped on the current image, super-imposing color signals from different phases of the cardiac cycle and reducing the impression of pulsa-tion. Low persistence settings should be selected for the heart, with higher persistence settings best for small vessels.

Gain demonstrates the percentage of color exhibited on the screen, similar to the gray-scale gain function. Artifacts occur when the gain is too high (Figure 2). However, gain settings in the mid-range may lead to superimposition of color over the border of a structure of interest, particularly when the AV valves are examined, giving the false impression of a septal defect[12] (Figure 2 left). The gain should initially be set to low and gradually increased until color information is optimal (Figure 2 right).

PLANES IN FETAL COLOR DOPPLER ECHOCARDIOGRAPHY

Fetal echocardiography includes assessment of the veno-atrial, AV and ventriculo-arterial connections. Different planes have been proposed to 'simplify' and standardize fetal cardiac examination, some adapted from neonatal cardiology and others specific for fetal conditions[13-21].

The four-chamber view (4CV) is the accepted standard plane in initial cardiac examination[22]; however, as less than 50% of all cardiac defects can be detected with the 4CV alone, it cannot be used in isolation[22,23]. The five-chamber view (5CV) is generally acknowledged to be the next most helpful plane, allowing visualization of the ascending aorta arising from the left ventricle (LV). The best plane(s) to visualize the pulmonary trunk, ductus arteriosus (DA) and aortic arch are debated[13-21], and include the short-axis view, the sagittal aortic arch and ductal arch views, and the three-vessel view (3VV)[13-21]. The latter allows optimal assessment of the pulmonary artery–DA and ascending aorta–aortic arch-isthmus pathways, the superior vena cava (SVC)[13,15,17,19], the relationship of these vessels to the trachea[19] and the presence of the thymus[21,24].

Color Doppler adds hemodynamic information gained throughout the cardiac cycle to morphological imaging[24]. The same planes for cross-sectional echocardiography can be used in assessment of flow dynamics during systole and diastole. Optimal color Doppler information can be obtained on these cross-sectional planes by using an insonation angle parallel to the flow events of interest. Recently Yagel *et al.*[19] proposed in this journal to achieve the comprehensive examination of the fetal heart by five short-axis views. In this review we postulate that three planes are sufficient to achieve reliable fetal color Doppler echocardiography: These are the 4CV, 5CV and 3VV (Figures 3–5).

The four-chamber view (4CV)

This transverse, cross-sectional plane enables simultane-ous demonstration of the left and right atria and ventricles, the interatrial and interventricular septa, the former being separated by the foramen ovale, and a transverse sec-tion of the descending aorta. Under good conditions, the pulmonary veins can be recognized draining to the left
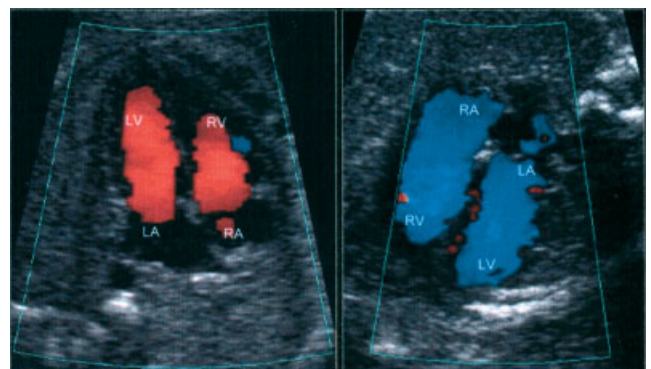

Figure 2 Gain. The same four-chamber view is shown with different gain values. The left image demonstrates a higher gain than necessary (here, 60%) leading to 'overlapping' of color across the septum, and an apparent ventricular septal defect. The image on the right shows use of an appropriate gain (50%), demonstrating separate atrioventricular valve perfusion (RA, LA, right and left atrium; RV, LV, right and left ventricle).

Figure 3 The four-chamber view is shown in diastole from the apical approach (left image), with blood flow toward the transducer in red, and from the basal approach (right image), with blood flow away from the transducer in blue. Two separate color stripes are evident, demonstrating blood flow from the atria into the ventricles. Compare the left image with the abnormal findings in Figures 6 and 7 (RA, LA, right and left atrium; RV, LV, right and left ventricle).
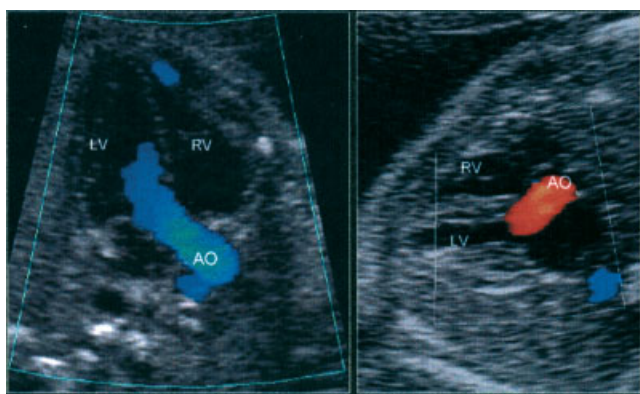
Figure 4 The five-chamber view is demonstrated in systole. According to fetal position, blood flow can be visualized either from the apical or left side (left image) with blood flow away from the transducer in blue, or from the right side (right image) (blood flow toward the transducer in red). The selection of an appropriately high velocity range demonstrates laminar non-aliased flow (AO, aorta; RV, LV, right and left ventricle).
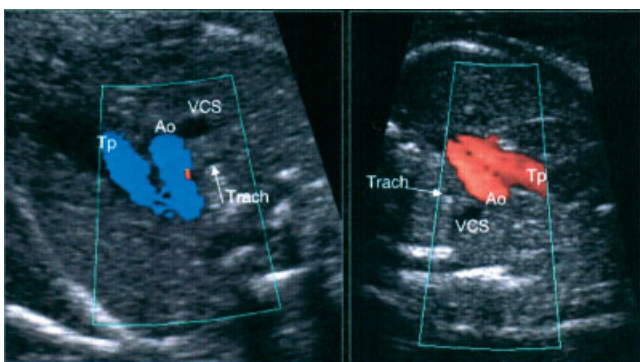
Figure 5 The three-vessel view seen in systole. In dorso-posterior position (left image) color Doppler imaging is obtained from the apical approach, demonstrating high velocity scale perfusion through the pulmonary trunk (Tp) and ductus arteriosus as well as the aortic arch (Ao) and isthmus (blood flow away from the transducer indicated in blue). Compare this image with the abnormal findings in Figures 10 and 11. In the dorso-anterior position (right image) blood streams towards the transducer in red. Both vessels appear on the left side of the spine and the trachea (Trach) is seen on the right posterior to the superior vena cava (VCS).

atrium (LA). The mitral and tricuspid valves can be visualized open during diastole and closed during systole. Use of cine-loop and magnification with the zoom function allows appreciation of valve excursion and myocardial contractility during the complete cardiac cycle. This is important information, but is more reliably and simply assessed by visualization of diastolic perfusion across the AV valves and lack of regurgitation during systole.
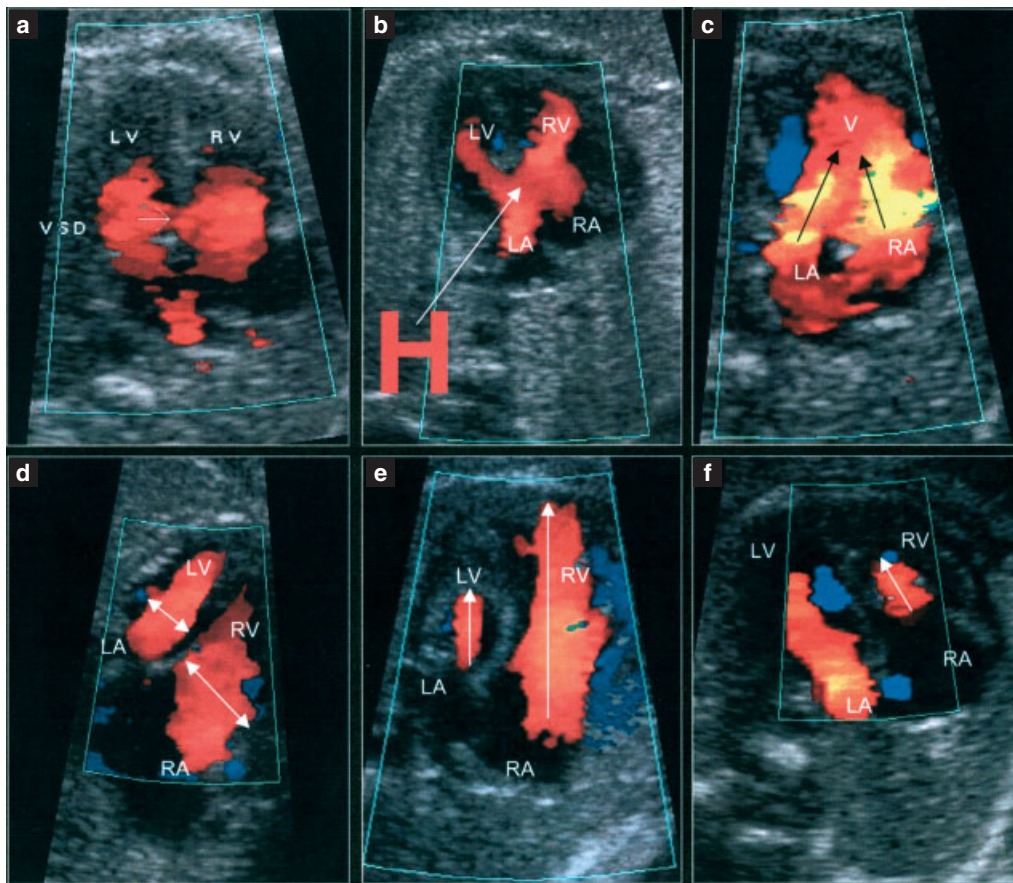
Diastole

Color Doppler examination should be performed from an apical or basal approach, with blood streaming toward or away from the transducer. If the heart is analyzed from the right or left side, the transducer should be shifted on the maternal abdomen to obtain a cross-section with

flow direction near the insonation angle with a range of $\pm 45°$. Diastolic perfusion from both atria into the ventricles across the AV valves can then be easily assessed and typically shows two red (apical) or blue (basal) stripes of equal size separated by the interventricular septum[11] (Figure 3).

Perfusion abnormalities are easily demonstrated in this manner, either with an abnormal pattern of bilateral perfusion (Figure 6) or with unilateral perfusion (Figure 7).

Visualization of two color stripes (Figure 6)

1 In *ventricular septal defect (VSD)* there is shunting across the interventricular septum, usually involving the muscular part of the septum, and rarely the inlet alone (Figure 6a). Most of these defects are not appreciated on real-time imaging and are incidentally detected with color Doppler examination. Bidirectional shunting is seen in isolated VSD[25]. Most VSDs, however, are not detected in this plane but in the 5CV.

2 In *atrioventricular septal defect (AVSD)* there is involvement of the lower part of the interatrial septum (septum primum), the upper part of the inlet ventricular septum, and the septal AV valves to a variable extent[26], leading to mixture of flow at the crux of the heart. Blood flow from the atria is not only directed into the corresponding ventricle but crosses the defect to reach the contralateral ventricle. This biventricular diastolic flow, communicating at the level of the AV valves, has a characteristic 'H' shape[10] (Figure 6b).

3 *Double-inlet ventricle with patent AV valves.* Two atria and two patent AV valves are seen overriding the ventricle. The diastolic blood flow commonly appears as two distinct color streams merging within the ventricle (Figure 6c).

4 *Disproportionate left and right ventricular color stripes* are typically found in coarctation of the aorta. There is reduction in left ventricular filling in comparison to the dilated right side (Figure 6d). When a VSD is found in association with LV:RV disproportion, other conditions with great vessel involvement should be considered, particularly double-outlet right ventricle (RV), tubular aortic arch hypoplasia or interruption of the aortic arch.

5 *Diminutive LV.* In some forms of hypoplastic left heart syndrome (HLHS) and late stages of critical aortic stenosis with left ventricular dysfunction, biventricular filling with a small left stripe may be seen, indicating patency of the mitral valve (Figure 6e). Blood leaves the ventricle either by mitral valve regurgitation (MR) or across the stenotic aortic valve. Rarely, ventriculo-coronary communications allow communication with the coronary system[27].

6 *Diminutive RV.* In some cases of pulmonary atresia with intact ventricular septum or in critical pulmonary stenosis there may be minimal perfusion of the right ventricular cavity (Figure 6f). Blood flowing into the diminutive RV may regurgitate back across

Figure 6 Heart anomalies with two color stripes in diastole in the four-chamber view (compare with Figure 3 left). Two stripes with a connection are seen in: (a) ventricular septal defect, (b) atrioventricular septal defect with the 'H' sign, or in (c) double-inlet ventricle with two atrioventricular valves but no septum. Two stripes with differing chamber size are seen in coarctation of the aorta (LV:RV disproportion) (d), some forms of critical aortic stenosis or hypoplastic left heart syndrome with diminutive left ventricle (e), or in hypoplastic right ventricle in some forms of pulmonary atresia (f) (RA, LA, right and left atrium; RV, LV, right and left ventricle; V, ventricle; VSD, ventricular septal defect).

the tricuspid valve or may enter ventriculo-coronary fistulae, which are a relatively frequent finding in such cases when ventricular filling is shown by color Doppler[28].

Visualization of one color stripe (Figure 7)

1 *HLHS.* Most prenatally-detected cases of HLHS show absent perfusion of the left ventricular cavity, characteristically with a single right-sided color stripe (Figure 7a and b).
2 *Mitral atresia with VSD.* A similar pattern to HLHS is demonstrated on color Doppler, but there is filling of the left ventricular cavity via a VSD in late diastole.
3 *Pulmonary atresia with intact ventricular septum.* Findings are as observed in HLHS, but the single color stripe is on the left side, and absent on the right (Figure 7d).
4 *Tricuspid atresia with VSD.* A unilateral left-sided color stripe is visualized, with later filling of the diminutive RV via a VSD (Figure 7e).
5 *Large AVSDs* may be associated with one (wide) stripe connecting the common atrial chamber with the ventricles via a large VSD (Figure 7f).

6 *Double-inlet ventricle with one atretic valve.* The two atria override the ventricular cavity, but only one AV valve is patent, giving a single large inflow stripe on color Doppler imaging (Figure 7c).

Systole

During systole both the mitral and tricuspid valves should be closed. Regurgitation of AV valves is suspected when blood flows from the ventricle into the atrium during systole. Atrial enlargement should prompt suspicion of insufficiency of the corresponding AV valve (Figure 8c). However, most cases of tricuspid and/or mitral valve regurgitation are detected by the routine application of color Doppler[3] (Figure 8a,b and d–f). Tricuspid valve regurgitation (TR) occurs not infrequently, in a trivial form (Figure 8a) in 3–5% of mid-trimester pregnancies[29,30]. Such trivial TR is usually evident during early systole, with a peak velocity not exceeding 200 cm/s, and resolves in most cases before term[29,30]. However, TR may indicate a range of other cardiac or extracardiac abnormalities and a checklist of conditions should be ruled out before assuming it is trivial[11]. Heart defects characterized by severe tricuspid valve
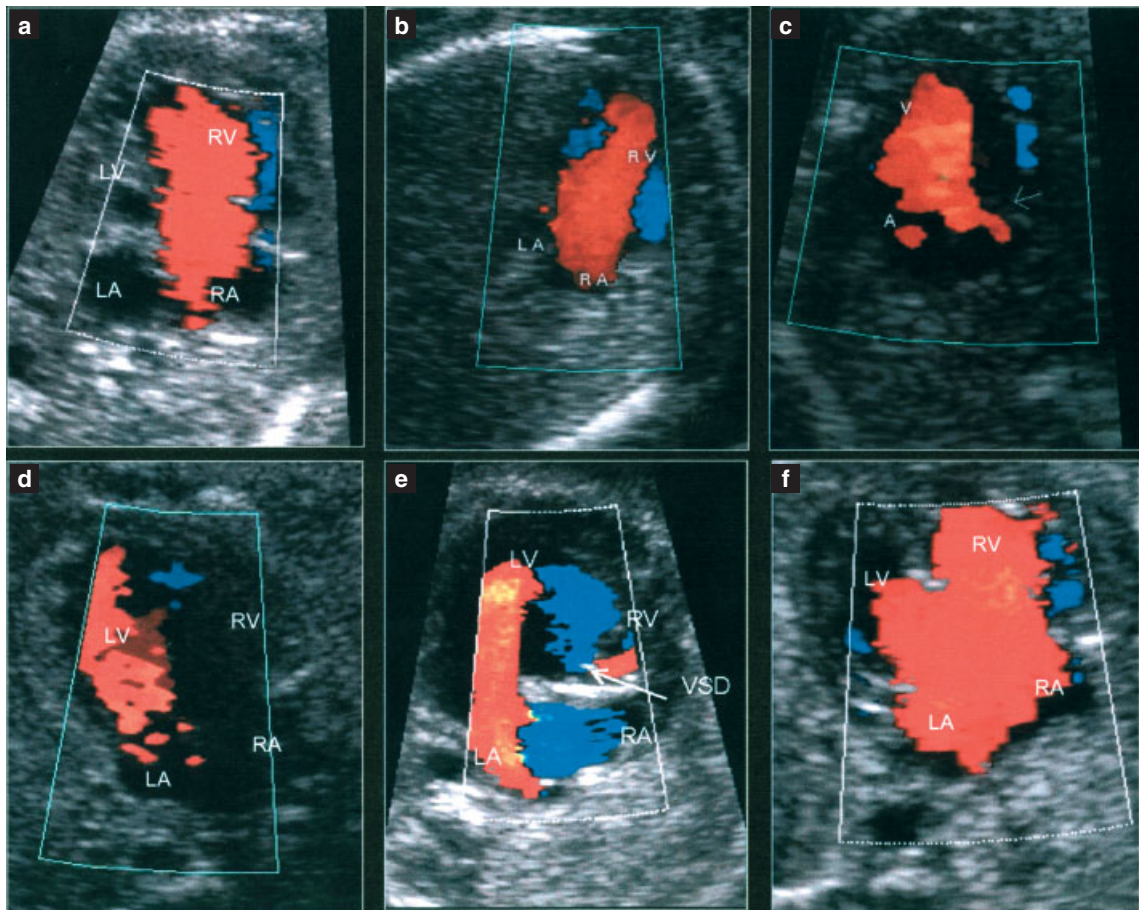
Figure 7 Heart anomalies with one color stripe in diastole in the four-chamber view (compare with Figure 3 left). Unilateral perfusion from the right atrium into the right ventricle in hypoplastic (a) or absent left ventricle (b). In example (c) there is a dextrocardia with single atrioventricular valve connecting into an indeterminate chamber in a fetus with right atrial isomerism (heterotaxy syndrome). In examples (d) and (e) there is a unilateral perfusion from the left atrium into the left ventricle due to a hypoplastic right ventricle: in (d) in pulmonary atresia and in (e) in tricuspid atresia with ventricular septal defect (VSD). In the latter the right ventricle is filled in late diastole via the VSD. Example (f) demonstrates a wide stripe in a large atrioventricular septal defect (A, atrium; RA, LA, right and left atrium; RV, LV, right and left ventricle; V, ventricle; VSD, ventricular septal defect).

insufficiency include Ebstein's anomaly and tricuspid valve dysplasia[31,32]. Both defects are often associated with marked dilatation of the right atrium (RA)[32] (Figure 8c). Of note, as the septal attachment of the tricuspid valve is lower than normal within the RV in Ebstein's anomaly, the regurgitant jet arises within the ventricle itself rather than at the usual AV valve level. Pulmonary atresia and pulmonary stenosis may also be associated with tricuspid valve insufficiency (Figure 8b) as well as an acquired constriction of DA (spontaneous or drug-induced).

Heart defects characterized by minor AV valvular regurgitation include AVSDs due to malformed valves[26] (Figure 8d), some cases of HLHS and aortic coarctation, because of compensatory right heart dilatation due to volume overload, and complex cardiac malformations with a single AV valve.

Mitral valve insufficiency is rarely present in cardiac defects but is characteristically found in aortic atresia or critical aortic stenosis with patent mitral valve. Clefting of the mitral valve found in septum primum defect (ASD I) may cause MR.

TR is associated with a range of conditions besides cardiac structural anomalies, including increased right ventricular afterload, volume overload and cardiomyopathy.

Concurrent MR and TR should prompt investigation for a systemic cause. Typical cardiac anomalies include primary dilated cardiomyopathy or cardiomyopathy secondary to maternal lupus erythematosus or to volume overload, present in tachycardia, arteriovenous fistula, anemia, or recipient in twin–twin transfusion (Figure 8f). Infective myocarditis, particularly cytomegalovirus infection (Figure 8e), should be excluded if additional signs are present.

Causes of tricuspid regurgitation are presented in Table 2.

The five-chamber view (5CV)

This plane is characterized by the aortic root emerging from the LV and the interventricular septum in continuity with the anterior wall of the ascending aorta. The ascending aorta courses slightly to the right as it forms the
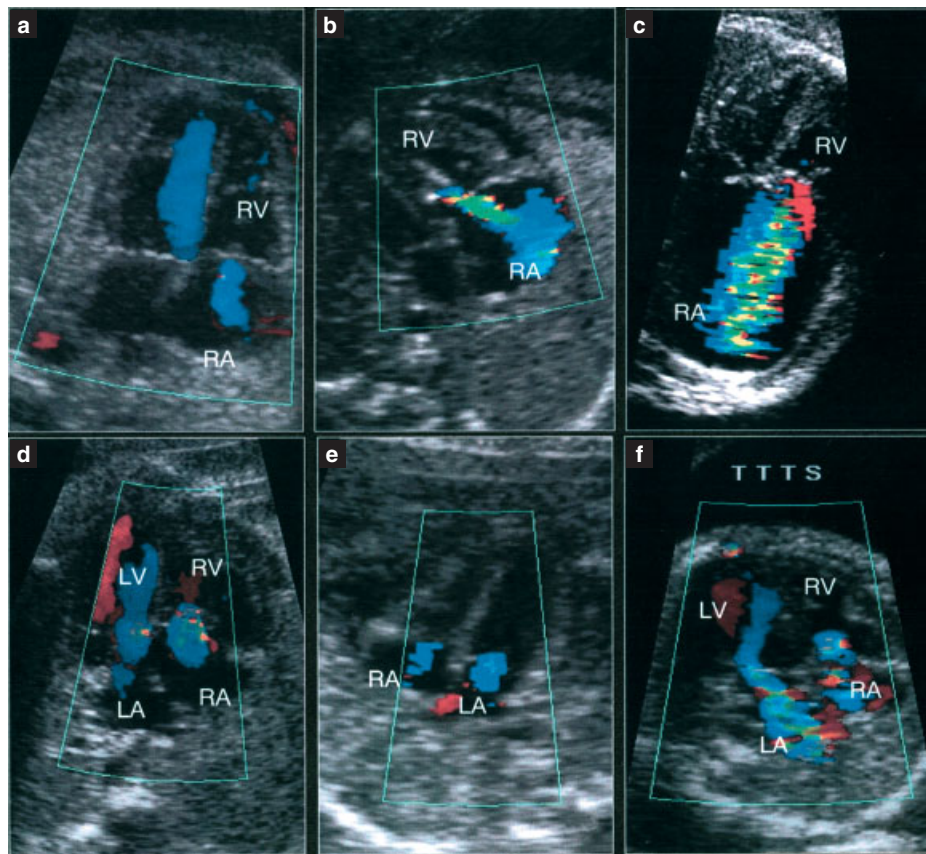
Figure 8 Atrioventricular valve insufficiency during systole. (a) Trivial tricuspid regurgitation (TR) from the right ventricle into the right atrium (RA). (b) TR in pulmonary atresia, which showed holosystolic high velocities on spectral Doppler. (c) Severe TR with cardiomegaly in tricuspid dysplasia and pulmonary atresia. (d) Bilateral regurgitation in a fetus with atrioventricular septal defect. (e) Bilateral regurgitation in a fetus with cytomegalovirus infection and myocarditis. (f) Bilateral regurgitation in the recipient in twin–twin-transfusion syndrome (RA, LA, right and left atrium; RV, LV, right and left ventricle).

Table 2 Causes of tricuspid regurgitation (adapted from Chaoui[11])

Pathogenesis	*Etiology*
Mild (trivial) TR	'Physiological' 3–5% of all pregnancies, and detected at 18–24 weeks' gestation, resolves during pregnancy
Heart defects with dysplasia of the TV	Ebstein's anomaly TV dysplasia
Heart defects and diseases with right ventricular outflow tract obstruction	Pulmonary atresia Pulmonary stenosis Constriction of the DA (mild to severe TR)
Heart defects with 'facultative' TR	Atrioventricular septal defect Hypoplastic left heart syndrome Coarctation of the aorta Double outlet RV
Volume overload (sometimes with MR)	Fetal anemia (Rhesus immunization, parvovirus infection) Peripheral arteriovenous fistula (vein of Galen aneurysm, sacrococcygeal teratoma, chorioangioma) Severe arrhythmia (tachycardia, bradycardia) Recipient in TTTS Heart defect with volume overload or pulmonary regurgitation (tetralogy of Fallot with absent pulmonary valve)
Impaired myocardial contractility (sometimes with MR)	Myocarditis: infection, autoimmune diseases (e.g. SLE) Cardiomyopathy: secondary to severe arrhythmia or volume overload, dilated cardiomyopathy Myocardial impairment in hypoxia: severe IUGR with increased peripheral vascular resistance Endocardial fibroelastosis

DA, ductus arteriosus; IUGR, intrauterine growth restriction; MR, mitral valve regurgitation; RV, right ventricle; SLE, systemic lupus erythematosus; TR, tricuspid valve regurgitation; TTTS, twin–twin transfusion syndrome; TV, tricuspid valve.

aortic arch (Figure 4). The laminar aortic flow appears red on color Doppler imaging during systole when scanned from the fetal right side (Figure 4 right), or blue with an apical or left-sided approach (Figure 4 left). Scale velocities should ideally be as high as possible (> 60 cm/s) since a lower velocity range will result in aliasing, mimicking turbulent flow.
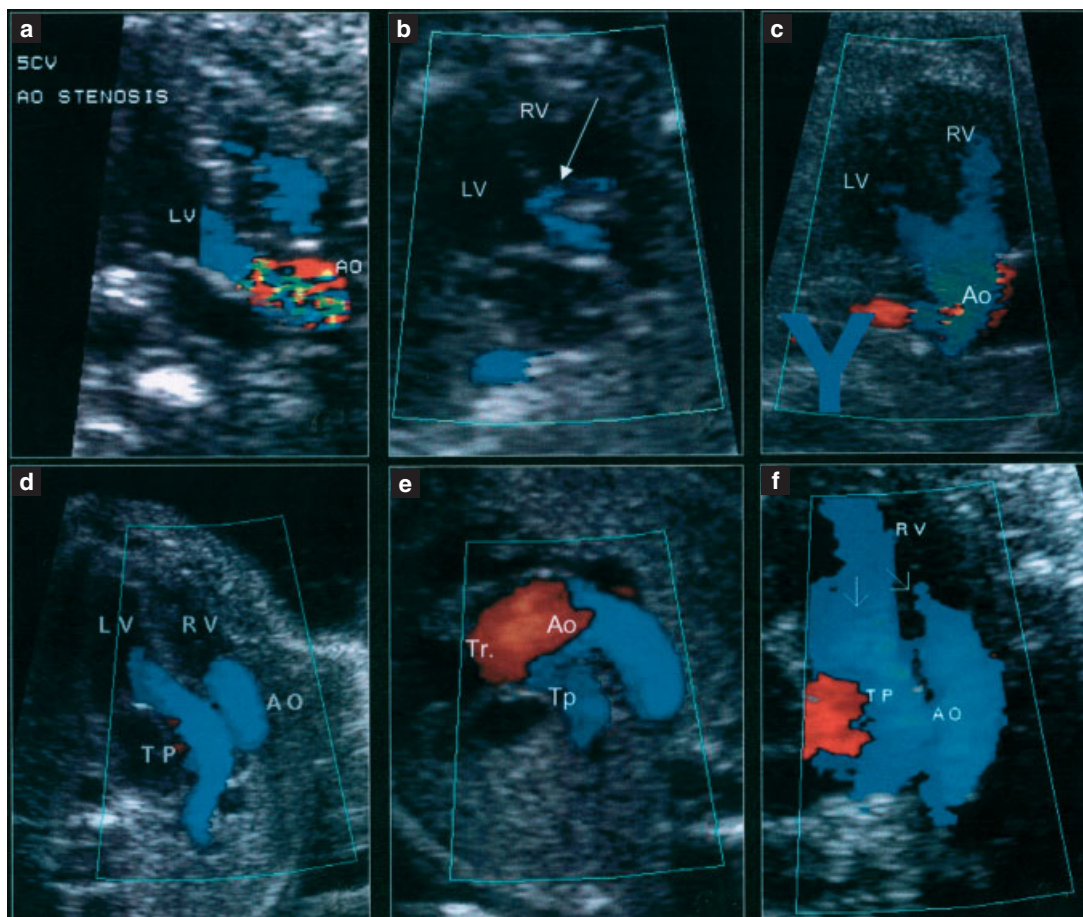
Systole

Despite an apparently normal 4CV, four abnormal situations can be identified on the 5CV with color Doppler (Figure 9).

1 Turbulent flow across the aortic valve in simple aortic stenosis[8] (Figure 9a).
2 Shunting through a perimembranous VSD with a normal aortic connection (Figure 9b).
3 Visualization of the origin of the pulmonary trunk in d-transposition of the great vessels, identified by its bifurcation into the main pulmonary arteries (Figure 9d).

4 Overriding of the aorta over both ventricles, connected by a VSD (Figure 9c).

Of these entities, the latter may indicate any of a number of possible cardiac defects, where final diagnosis is made by evaluation of the pulmonary trunk and pulmonary arteries. An overriding aorta may give a characteristic 'Y' color Doppler sign (Figure 9c)[10].

i *Malalignment VSD with overriding aorta* is suspected when there is a patent pulmonary artery with regular flow. This is rare, but has been described as a typical heart anomaly in fetuses with trisomy 18[20].
ii In *tetralogy of Fallot* there is associated pulmonary stenosis. The overriding aorta is generally dilated and the pulmonary trunk is variably small with a patent pulmonary valve; blood flow in the pulmonary trunk is not necessarily turbulent and pulsed Doppler velocities > 180 cm/s are rarely found.
iii *Pulmonary atresia with VSD* is associated either with a tiny or diminutive pulmonary trunk and a closed pulmonary valve. Occasionally there is even absence of the pulmonary trunk. Retrograde flow across the

Figure 9 Heart anomalies detectable on the five-chamber view during systole (compare with normal findings in Figure 4 left). (a) Turbulent flow across the stenotic aortic valve. (b) Small ventricular septal defect (VSD) with shunting into left ventricle during systole. (c) VSD with an overriding aorta, with perfusion from both ventricles into the ascending aorta suggesting the letter 'Y'. (d) d-Transposition of the great arteries with discordant connections of the great vessels. (e) Truncus arteriosus communis Type 1 with the truncus (Tr) bifurcating into the aorta (Ao) and pulmonary trunk (Tp). (f) Double-outlet right ventricle with both vessels arising from the right ventricle. RV, LV, right and left ventricle.

DA to minute right and left pulmonary arteries is visualized on color Doppler.

iv *Absent pulmonary valve syndrome* occurs with severely dysplastic or absent valve leaflets with stenosis and insufficiency. There is often associated absence of the DA, right-sided aortic arch and marked dilatation of the pulmonary arteries (> 10 mm in diameter in mid-gestation).

v *Truncus arteriosus communis Type I* is characterized by a common arterial trunk giving rise to the pulmonary trunk and aorta, demonstrated by color Doppler (Figure 9e).

Diastole

The aortic valve should be closed and non-regurgitant in diastole. Aortic root regurgitation is extremely rare and usually suggests valve dysplasia[8], sometimes found in truncus arteriosus communis or combined with aortic stenosis[33,34]. However, severe dilatation of the aortic root and aortic regurgitation has also been described prenatally in a case of Marfan syndrome[31]. In early and mid-pregnancy, an absent or dysplastic aortic valve may show to-and-fro blood flow[33,35] with similar features to absent pulmonary valves; however, most cases become hydropic[35], evident on nuchal translucency screening[33,36], or die *in utero*[33,35].

The three-vessel view (3VV)

This plane is obtained at the level of the fetal mediastinum by moving the transducer obliquely cephalad from the 4CV[13,15,17,19,21]. The pulmonary trunk, DA, aortic arch, aortic isthmus and SVC are demonstrated, with the aortic and ductal arches forming a V-configuration pointing to the posterior spine on the left (Figure 5)[37]. The SVC lies to the right of the aortic arch. The trachea may be identified as a bright-walled structure lying to the right of the great vessels and posterior to the SVC, thus the coined term, '3VT' view, for 'three vessels and trachea'[19]. The pulmonary view has been deliberately omitted in this review as it is our opinion that most significant anomalies can be identified in the color Doppler 3VV. With abnormal findings, however, separate visualization of the bifurcating right and left pulmonary arteries may be important. Many anomalies of the outflow tracts can be identified in this transverse upper-thoracic plane, which is achievable in most fetal positions[11,21]. Color Doppler information is easily obtained when the vascular structures of interest are not perpendicular to the angle of insonation. The convergence of the vessels at the level of the isthmus and DA is characteristically 'V'-shaped, colored blue when the fetal spine is posterior (Figure 5 left) and red when it is anterior[8,9] (Figure 5 right).

In the 3VV the following should be evident:

1 Aorta and pulmonary trunk converging toward the left thorax with the trachea to their right.

2 The pulmonary trunk with a slightly greater caliber than the aorta (ratio 1.2 : 1).
3 The straight course of the vessels.
4 Antegrade flow through both great vessels throughout the cardiac cycle.

In a plane more cranial to these vessels and just behind the thymus the innominate vein can be seen connecting the left jugular vein and the SVC. Its course is transverse and parallel to the sternum with blood flow toward the right side and should not be misinterpreted as arising from the pulmonary artery or DA.

In the 3VV deviation from normal findings can demonstrate the findings described below.

Visualization of two antegrade color stripes (Figure 10)

1 *Small pulmonary artery with antegrade laminar flow* (Figure 10 left) occurs in pulmonary stenosis with shunting, either from VSD or TR. This pattern is observed in conditions such as Ebstein's anomaly, tetralogy of Fallot with mild pulmonary stenosis, double-outlet RV with pulmonary stenosis, or tricuspid atresia with VSD. Antegrade flow across the DA indicates a non-ductus dependent pulmonary circulation.

2 *Small aortic arch with antegrade flow* may represent coarctation of the aorta, particularly tubular hypoplasia. The aortic arch is tortuous or very small, and flow across the isthmus allows differentiation from interruption of the aortic arch. Of note, an apparently small aortic arch associated with a slightly dilated pulmonary trunk can be a normal finding in third-trimester fetuses (> 32 weeks).

3 *Dilated pulmonary artery with turbulent antegrade flow* can be found in isolated pulmonary stenosis[8]. Aliasing occurs in the region of the pulmonary valve with poststenotic dilatation. Rarely, in severe cases there may be reversed flow across the ductus, indicating possible duct-dependence in the neonatal period[37].

4 *Dilated aortic arch with antegrade turbulent flow* can be seen in isolated aortic stenosis without left ventricular dysfunction. Poststenotic dilation is typical, but the diagnosis should have been suspected in the 5CV.

5 *Right-sided aortic arch* (Figure 10 center) should be suspected when the trachea is not present on the right of both vessels but lies either between them or, rarely, to their left. In the former, the aorta courses to the right of the spine and the pulmonary trunk to the left with the trachea entrapped between them. The DA joins the aorta posterior to the trachea, giving the 'U'-sign.

6 *Constriction of the DA*. The DA sustains the highest velocities within the fetus and therefore normally exhibits aliased flow in the third trimester. With ductal constriction, there is continuous flow throughout the cardiac cycle and turbulent flow at the level of constriction[8], easily confirmed with pulsed or continuous wave Doppler.
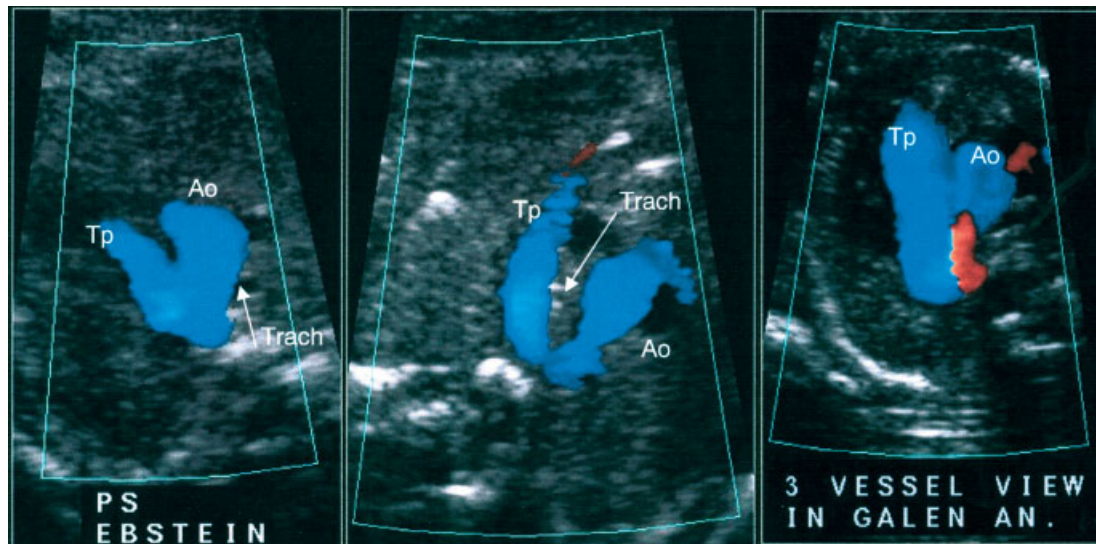
Figure 10 Three-vessel view with two antegrade flow stripes (compare with Figure 5). Tiny pulmonary trunk with antegrade perfusion in mild pulmonary stenosis (left), in Ebstein's anomaly in this case. Right-sided aortic arch (Ao) with the trachea (Trach) entrapped between the aortic arch and pulmonary trunk (Tp) (center). Retrograde perfusion across the isthmus due to increased cerebral perfusion, in a case of vein of Galen aneurysm (right).
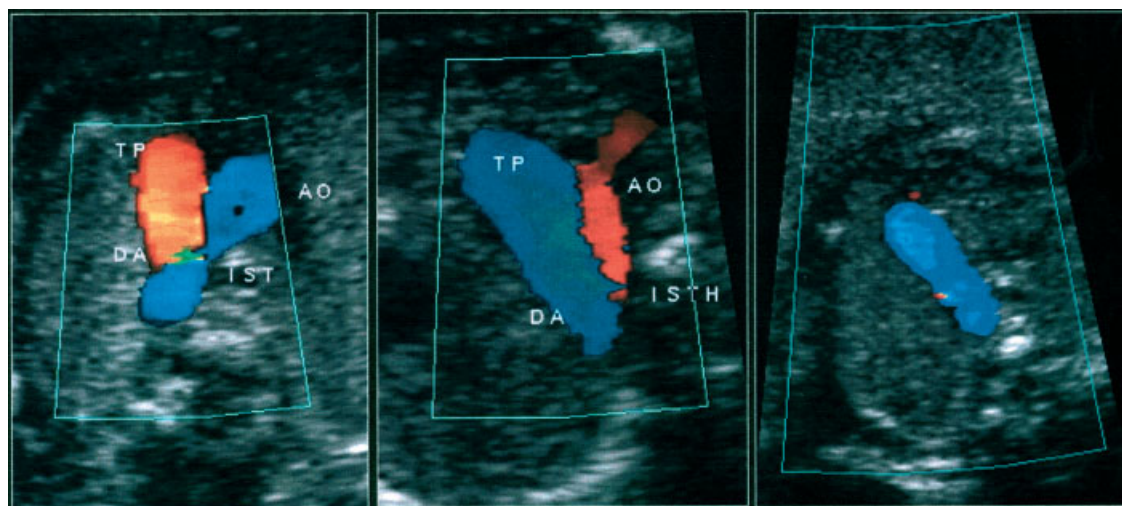
Figure 11 Three-vessel view with one antegrade flow stripe. In severe right ventricular outflow tract obstruction (left) such as in this case of pulmonary atresia, there is retrograde perfusion of the ductus arteriosus (DA) into the pulmonary trunk (TP). In severe left ventricular outflow tract obstruction (center), demonstrated here in a case of hypoplastic left heart syndrome, there is retrograde perfusion of the aortic arch (AO). In this fetus only one stripe is visualized (right). In such cases, the transducer should be turned to evaluate whether the other vessel is hypoplastic with retrograde flow, or whether the vessels are transposed (ISTH, aortic isthmus).

Visualization of one antegrade color stripe (Figure 11)

Occasionally one antegrade stripe only is identified. If the one vessel is of normal caliber (Figure 11 right), the examiner should turn the transducer 90° to establish whether both vessels are present with parallel course, but superposed. If the single vessel is dilated, however, it is likely that the other vessel is small or hypoplastic. Slight superior or inferior movement of the transducer may reveal the small and retrogradely-perfused vessel. However, late-diastolic retrograde perfusion may be seen as a normal finding within the aortic isthmus in the late third trimester or in compromised fetuses with intrauterine growth restriction, probably as a result of increased cerebral perfusion[38], and should not be mistaken for left heart obstruction.

1 *Antegrade flow in the aorta and retrograde flow in the pulmonary artery and DA* (Figure 11 left) occurs in pulmonary atresia or severe pulmonary stenosis. Pulmonary atresia, especially with an associated VSD, can lead to a small or even absent pulmonary trunk with a tortuous DA. The small, tortuous ductus may be visualized in a slightly lower plane than the usual 3VV, a helpful sign as the DA and pulmonary artery may not be easily identified in severe cases on real-time ultrasound.

2 *Antegrade flow in the pulmonary artery and DA and retrograde flow in a tiny aorta* (Figure 11 center) is seen

in aortic atresia or severe aortic stenosis. Severe aortic stenosis is usually associated with a straight aortic arch of small caliber. In aortic atresia, the aortic arch may be difficult to identify on real-time ultrasound, and is very small and tortuous.

3 *Transposition of the great arteries, double-outlet RV and other malpositioning anomalies of the vessels.* A normal 3VV is rarely present in these conditions. Usually one vessel is visualized and the other vessel may be seen while turning the transducer longitudinally. With associated pulmonary stenosis or tubular hypoplasia of the aorta, color Doppler may facilitate appreciation of difference in caliber.

4 *Interruption of the aortic arch* is a difficult diagnosis on real-time and color Doppler ultrasound. In the 3VV, the pulmonary trunk–DA pathway only is completely visualized. The ascending portion only of the aorta is seen and there is no connection with the DA.

5 *Venous anomalies in the 3VV.* The azygos vein joins the SVC in this plane, but flow within the vein normally cannot be seen unless there is increased perfusion, such as in azygos continuity with interruption of the inferior vena cava in left isomerism (polysplenia). Increased flow within the SVC may occur as a result of intracranial arteriovenous fistulae, e.g. vein of Galen aneurysm. In such cases retrograde flow within the isthmus permits obligatory high perfusion of the brain.

If an abnormal fourth vessel is seen on the left of the pulmonary trunk this indicates persistence of the left SVC. Diagnosis is aided by identification of a dilated coronary sinus at the level of the heart and visualization of the vein at the LA. As 3VV assessment of the pulmonary trunk and aorta are performed at high-velocity settings, flow within the SVC may not be appreciated.

CONCLUSIONS

It is our opinion that the three color Doppler echocardiographic planes presented here allow easy detection and precise description of the majority of significant fetal cardiac anomalies. We are aware that not all anomalies will be identified by this approach alone, but hope that the doubtful reader will be encouraged to use color Doppler imaging routinely in prenatal cardiac examination. Furthermore, recent experience shows that color Doppler is enormously helpful in early fetal echocardiography[39] at the time of the 11–14-week scan, where real-time examination of the heart is currently limited. Color Doppler examination between 13 and 14 weeks can demonstrate most of the findings presented in this review.

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