

# A Review of Findings in Fetal Cardiac Section Drawings

## Part 1: The 4-Chamber View

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**Objectives.** The goal of this presentation is to review some of the common and rare fetal heart abnormalities and to provide an easy approach to these findings with schematic drawings. **Methods.** Over the past 10 years, we collected cases in which the common views of the heart were abnormal and the differential diagnoses that existed for each. The presentation shows the normal sonographic sections and then variations of these sections and the associated anomalies. We used illustrative drawings to present these findings, enabling us to point out the main sonographic features of abnormalities of the heart. **Results.** The work reviews 17 fetal heart abnormalities in schematic drawings. **Conclusions.** This short review highlights several of the anomalies that can be recognized on the common sonographic views. The drawings tend to simplify the findings but serve as a basis for those doing fetal echocardiography when they encounter an unusual finding. **Key words:** drawings; fetal echocardiography; prenatal sonography.

**D**uring the past decade, several views have been suggested to show various cardiac anomalies.<sup>1-18</sup> Most people probably use the 4-chamber view, the 3-vessel-trachea view, outflow tracts, and, when appropriate, additional views, yet there is not an easy reference for the person who reads an examination to have an “atlas” of the abnormal findings in these views. The purpose of this 3-part series is to put together common anomalies that can be recognized in some particular locations during ultrasound investigation.

To avoid confusion in the reading of an image, we purposefully excluded sonograms and simply used drawings. Drawings allow the reader to unquestionably recognize the abnormal from the normal, and they allow “simplification” of the anatomy by merging in a single image structures that are on adjacent planes, such as the stomach and the heart.

In this first part of our 3-part series, we will discuss the anomalies visible at the level of the 4-chamber view of the heart.

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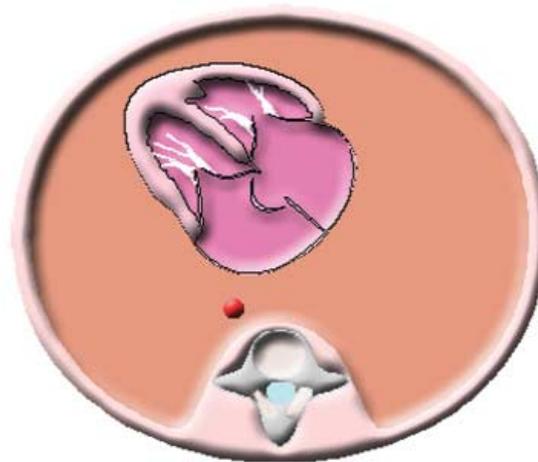
## Materials and Methods

Over the past 10 years, we collected cases in which the common views of the heart were abnormal and the differential diagnoses that existed for each. Some diagnoses are quite uncommon and unlikely to be found in regular practice, but they present such typical images that they are worth knowing.

The colors used in the drawings are conventional medical artist colors, with arteries in red and veins in purple or blue. The colors do not represent flow directions, as in color Doppler sonography, for instance, and do not correlate with fetal blood oxygenation. The sections will all be presented with the fetus in a cephalic dorsal position (Figure 1). Although this does not represent the conventional position used in computed tomography and magnetic resonance imaging, it will be more familiar to those who do prenatal ultrasound examinations.

The first image is a reference drawing of the basic normal 4-chamber view (Figure 2), with the cardiac apex and the descending aorta on the left and the axis of the heart at about 45° from the midline. The drawings are simplified; thus, small details such as the positions of valve leaflets are not depicted precisely.

**Figure 1.** Fetus in the cephalic dorsal position. The subsequent images are drawings of abdominal and thoracic sonographic views depicted as if the fetus were in this position.



**Figure 2.** Basic normal transverse thoracic view for comparison. The normal position of the heart and aorta (red) at the level of 4-chamber view is shown. This image serves as a reference for comparison with the following images showing anomalies.

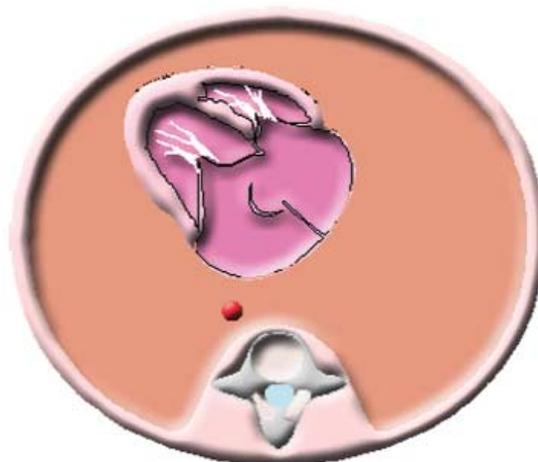
## Results

### Case 1

In this case (Figure 3), there is asymmetry between the sizes of the left and right ventricles.<sup>19,20</sup>

To determine possible causes, first, let us assume that this is a decreased right ventricle. The causes would include a hypoplastic right ventricle, as in pulmonary atresia with an intact ventricular septum or with a ventricular septal defect, early severe pulmonary stenosis, and early tricuspid stenosis. Early pulmonary stenosis is a possible cause because when the stenosis

**Figure 3.** Case 1. Four-chamber view showing asymmetry of ventricular size that could represent either a small right ventricle or a dilated left ventricle.



develops early, the right ventricle does not develop, and the right heart becomes hypoplastic. If the stenosis occurs late (eg, after 20 weeks), then the right ventricle has already developed, and the tricuspid valve becomes incompetent, the blood regurgitates, and the right atrium enlarges. We will see that the same mechanism occurs on the left side, too. To a certain extent, the same occurs with the atrioventricular valves as well.

Instead of a decreased right ventricle, this could also correspond to an increased left ventricle. This would probably be due to late aortic stenosis or mitral regurgitation.

#### Case 2

The dominant finding in this case is a small left ventricle (Figure 4). This will be associated with symmetric diagnoses of the first case. The possible causes include a hypoplastic left ventricle, as in hypoplastic left heart syndrome with aortic atresia,<sup>21</sup> early severe aortic stenosis or coarctation,<sup>18</sup> and early mitral stenosis or regurgitation, but also, there is an association with a left persistent superior vena cava, total anomalous pulmonary venous drainage, and premature closure of the ductus arteriosus<sup>22-24</sup> or foramen ovale.<sup>25,26</sup>

Conversely, case 2 can represent a large right ventricle, and mostly the symmetric diagnoses of the first case plus a few originals must be considered. This finding can represent late pulmonary stenosis and late tricuspid regurgitation, an absent pulmonary valve with pulmonary regur-

gitation, or a double-outlet right ventricle. The additional diagnoses include high-flow situations such as the recipient in twin-twin transfusion syndrome, a vein of Galen aneurysm,<sup>27-29</sup> chorioangioma of the placenta<sup>30-32</sup> when it exceeds 5 cm, and, rarely, hepatic angioma.<sup>33</sup>

#### Case 3

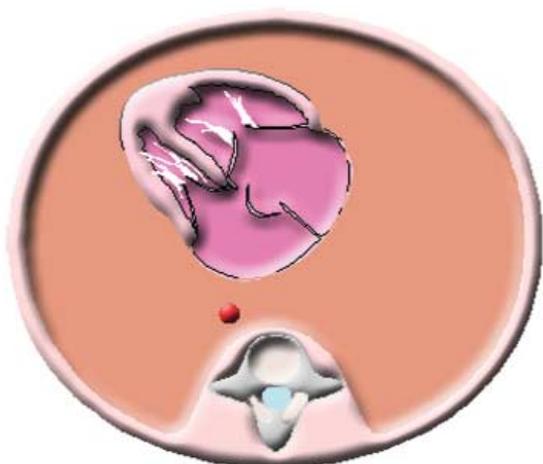
Case 3 (Figure 5) represents a left-axis deviation, in which the interventricular septum is almost at 90° degrees to the anteroposterior line.<sup>34,35</sup>

This finding is typical of conotruncal anomalies. (In many of these cases, the descending aorta is on the right side of the spine, as we will see in subsequent drawings.) The cardiac displacement can also be due to extracardiac anomalies, causing a shifting to the left by a right-sided anomaly such as a right-sided diaphragmatic hernia, a cystic adenomatoid malformation, and other developmental anomalies of the right lung, a right-sided pleural effusion, and other causes. In abdominal wall malformations such as gastroschisis and omphalocele, the heart may be shifted to the left as well.

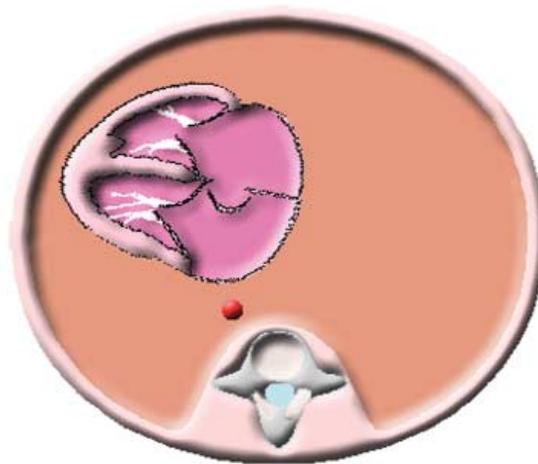
#### Case 4

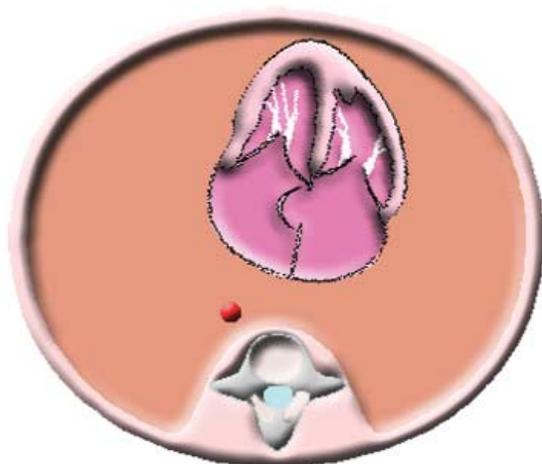
Case 4 represents a right-axis deviation (Figure 6), in which the interventricular septum is almost parallel to the anteroposterior line. There are more causes of right-axis deviation than left-axis deviation. They include a left-sided diaphragmatic hernia, isomerism and situs inversus, inversion of the ventricles (in congenitally cor-

**Figure 4.** Case 2. Four-chamber view showing asymmetry of ventricular size that could represent either a smaller left ventricle or a dilated right ventricle.



**Figure 5.** Case 3. Four-chamber view showing a left-axis deviation of the heart.





**Figure 6.** Case 4. Four-chamber view showing a right-axis deviation of the heart.

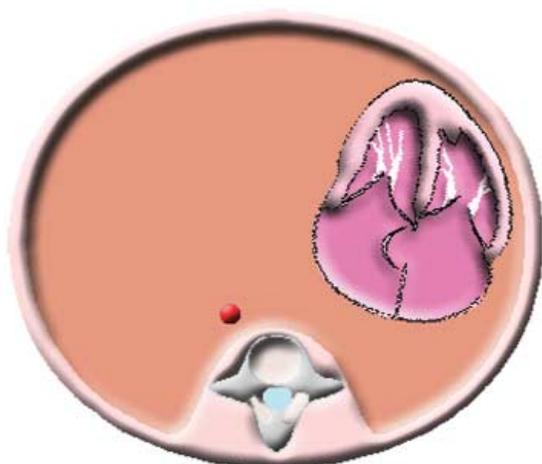
rected transposition or levotransposition), atrio-ventricular septal defects, a double-outlet right ventricle, and a common atrium.<sup>34,36</sup>

**Case 5**

Compared to the previous case, the heart is now totally along the right chest wall (Figure 7). This is called dextroposition.<sup>37</sup>

This may result from a contralateral diaphragmatic hernia, a cystic adenomatoid malformation of the lung, congenital lobar emphysema, a pleural effusion, chylothorax or ipsilateral lung agenesis, and hypoplasia. The latter can be associated with Scimitar syndrome. Several of these diagnoses would also be valid if the heart were shifted against the left side of the chest.

**Figure 7.** Case 5. Four-chamber view showing displacement of the heart to the right.



**Case 6**

In case 6, the heart looks “inverted” compared to normal (Figure 8). This phenomenon is called dextrocardia, and it results from a looping anomaly during embryologic development.<sup>38,39</sup>

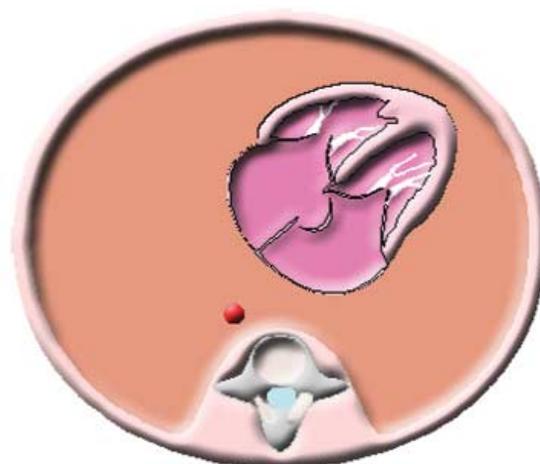
**Case 7**

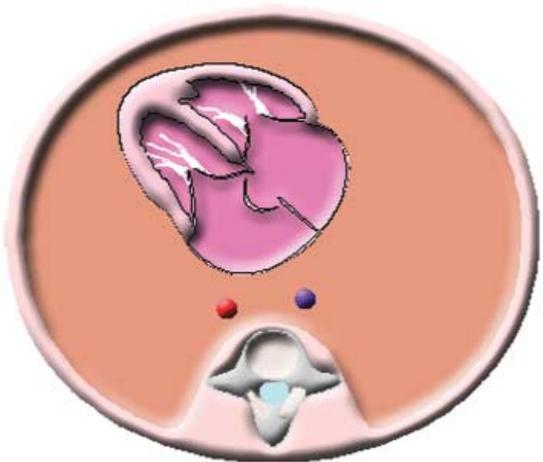
In case 7, there is a vein behind the 4-chamber view (Figure 9). It would be tempting to think this is the inferior vena cava, but at the level of the 4-chamber view, the inferior vena cava has already drained into the right atrium. This vessel represents the azygous or hemizygous continuation of an inferior vena cava interruption.<sup>40</sup> This finding can be isolated or a part of left isomerism, a condition called “polysplenia” in the past.

**Case 8**

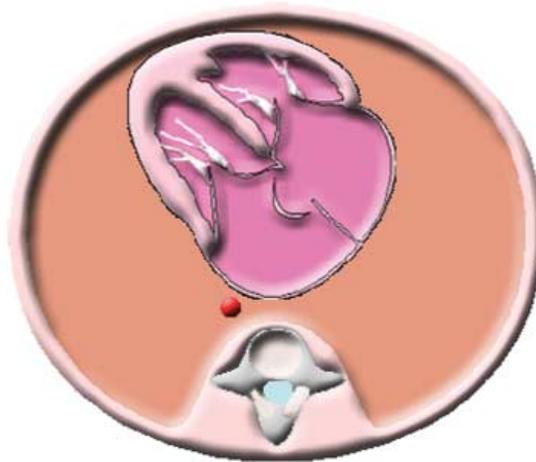
In case 8, the aorta is descending on the right of the spine (Figure 10). This may be seen in some conotruncal anomalies such as tetralogy of Fallot, truncus arteriosus, pulmonary atresia with a ventricular septal defect, and an absent pulmonary valve, but it could be an isolated sign in a fetus with a right-sided aortic arch.<sup>41,42</sup> One can have a right-descending aorta with a “left” or “right” aortic arch (terminology that refers to the position of the arch compared to the trachea, not the absolute position of the arch in the chest).

**Figure 8.** Case 6. Four-chamber view showing dextrocardia. The anatomic right ventricle and atrium are located on the left side, and the left ventricle and atrium are located on the right side, but the reverse could also be present. The axis of the heart is directed to the right.





**Figure 9.** Case 7. Additional vessel (purple) behind the 4-chamber view next to the aorta (red). This vessel represents an azygous or hemizygous continuation of an interrupted inferior vena cava.



**Figure 11.** Case 9. Four-chamber view showing global enlargement of the heart.

#### Case 9

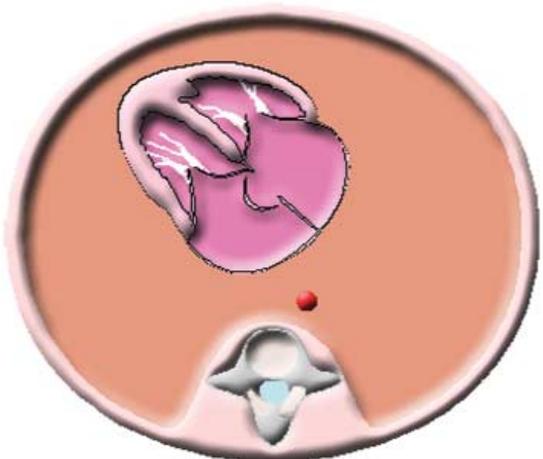
In case 9, the heart is globally enlarged (Figure 11). This can result from, or be present in, many different conditions,<sup>43</sup> such as endocardial fibroelastosis, fetal anemia,<sup>44</sup> storage disorders, ischemia,<sup>45</sup> infections,<sup>46,47</sup> atrioventricular and semilunar valve regurgitation after tachycardia, and an atrioventricular block. Some extreme forms of shunts,<sup>48</sup> such as a vein of Galen “aneurysm” (in fact a varix)<sup>49,50</sup> and chorioangioma, and obstructive lesions (aortic stenosis, idiopathic infantile aortic calcinosis, premature closure of the ductus, a true knot of the cord, and tumors in the outflow tracts) also cause global

enlargement. Finally, an apparent enlargement (with a normal heart but a too-small chest) may be found in severe intrauterine growth restriction, skeletal dysplasias, and severe oligohydramnios from absent renal function (renal agenesis and multicystic kidney dysplasia).

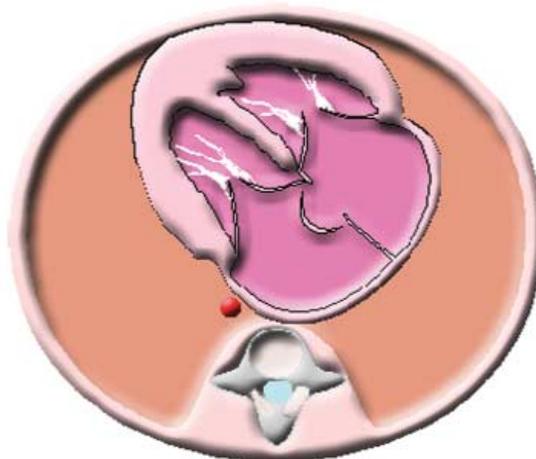
#### Case 10

In contrast, in case 10, the myocardium is enlarged (Figure 12), and there are just a few conditions that cause myocardial thickening. They include hypertrophic cardiomyopathies,<sup>51–53</sup> univentricular hearts (but the septum would be decreased or absent), and septal hypertrophy in

**Figure 10.** Case 8. Aorta descending on the right side of the spine at the level of the 4-chamber view.



**Figure 12.** Case 10. Thickened myocardium at the level of the 4-chamber view.



fetuses of diabetic patients (in which cases, the septum is predominantly affected).<sup>54,55</sup> In fetuses with bilateral renal anomalies and anhydramnios (in renal agenesis or multicystic kidneys), a thickened myocardium can be observed from 20 weeks onward.

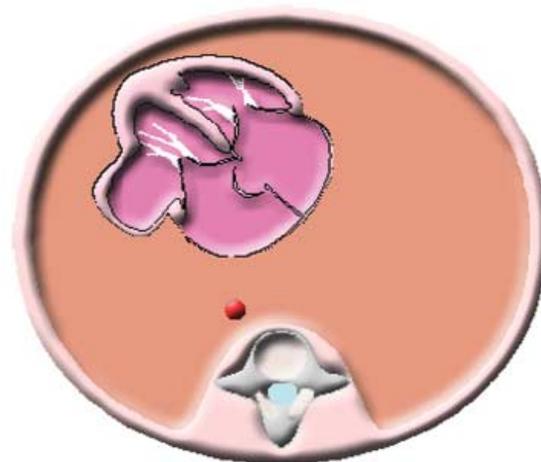
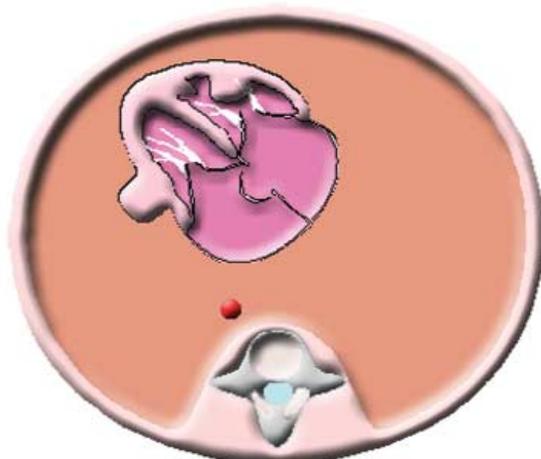
**Case 11**

Localized enlargement of the myocardium as in case 11 (Figure 13) may result from diabetic cardiomyopathy (and again mostly at the level of the septum) and, rarely, a myocardial infarct (in which case the affected region is usually initially hypoechoic), but predominantly this will represent a cardiac tumor such as rhabdomyoma.<sup>56</sup> Nodular hypertrophy may also present a similar image.<sup>57</sup>

**Case 12**

An “outpouching” of the myocardium such as in case 12 (Figure 14), may represent a cardiac diverticulum if the rest of the ventricular contractility is preserved or an aneurysm if the ventricle has little function left.<sup>58–60</sup> This distinction is easily made with color Doppler sonography. In a cardiac diverticulum, the ventricle will show flow, whereas in an aneurysm, little evidence of color will be seen, not only in the “outpouching” but also in the ipsilateral ventricle. Rarely, a pericardial cyst (which is more common in the atrioventricular groove) may give a similar appearance.<sup>61</sup>

**Figure 13.** Case 11. Localized enlargements of the left and right myocardia at the level of the 4-chamber view.



**Figure 14.** Case 12. Localized outpouching of the left ventricle at the level of the 4-chamber view.

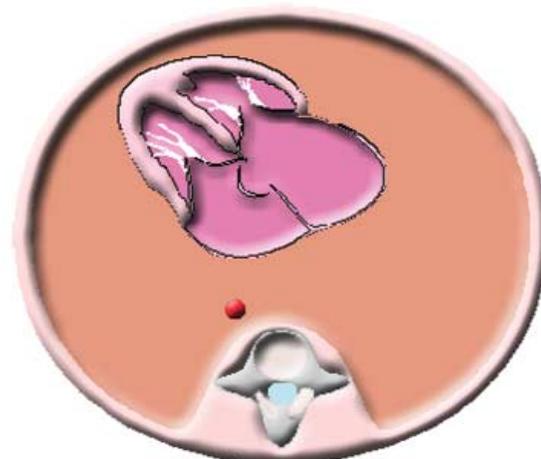
**Case 13**

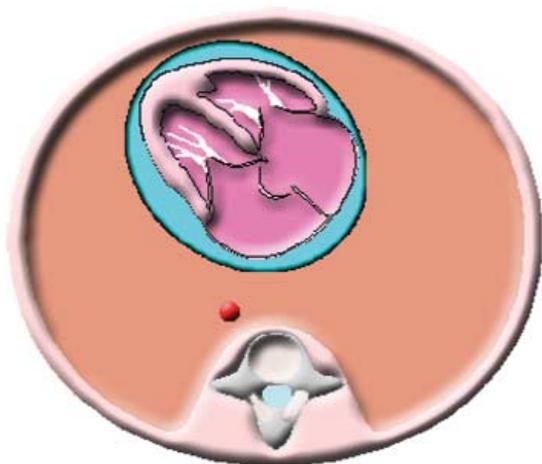
Case 13 represents an enlarged right atrium (Figure 15). This occurs in a variety of conditions such as tachyarrhythmias, Ebstein and Uhl anomalies, idiopathic right atrial enlargement, a common atrium, pulmonary atresia with a regurgitating tricuspid valve, pulmonary stenosis with an intact septum, and premature closure of the foramen ovale or ductus arteriosus.<sup>62</sup>

**Case 14**

A small amount of pericardial fluid or a small effusion, as in case 14 (Figure 16), can be present normally<sup>63</sup> or can be a sign of many conditions,

**Figure 15.** Case 13. Isolated enlargement of the right atrium at the level of the 4-chamber view.





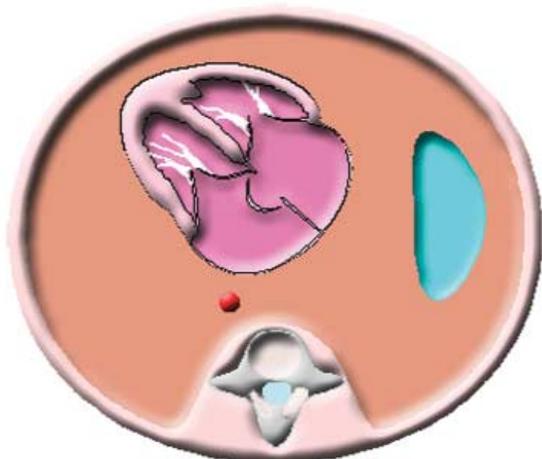
**Figure 16.** Case 14. Four-chamber view showing a pericardial fluid collection around the heart (cyan).

such as trisomy 21, a hypoplastic left heart, teratoma,<sup>64</sup> rhabdomyoma, hemangioma,<sup>65</sup> tachyarrhythmia, chorioangioma, sacrococcygeal teratoma,<sup>66</sup> an atrioventricular septal defect, cardiomyopathy, Rh disease, renal agenesis, posterior urethral valves, and twin-twin transfusion syndrome.<sup>67,68</sup>

**Case 15**

In case 15, the stomach is not on the same side as the heart (Figure 17). This may be called many

**Figure 17.** Case 15. Four-chamber view showing the normal position and structure of the heart. The stomach (cyan) is located on the opposite side of the heart (visceral situs). This image is simplified because the level of the stomach and the 4-chamber view of the heart are depicted at the same level, whereas in reality the stomach is lower in the fetus.



different names, such as heterotaxy, situs ambiguous, and visceral situs. This is a very practical observation because in all second-trimester obstetric ultrasound examinations, the 4-chamber view of the heart and the stomach are imaged. If they are not on the same side, there is a high risk (>95%) of cardiac anomalies.<sup>69,70</sup>

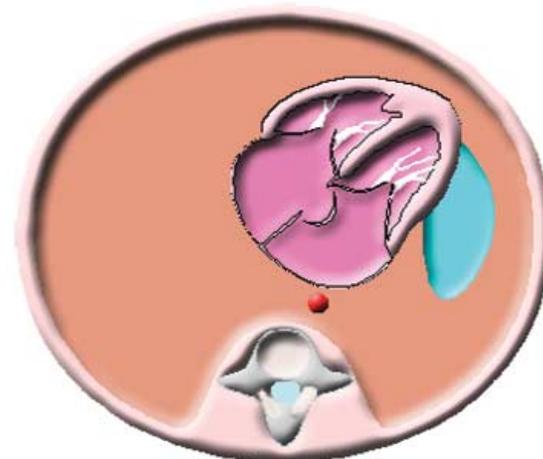
**Case 16**

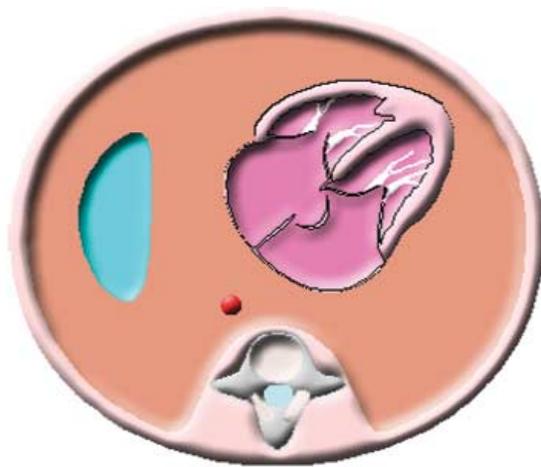
Compared to case 15, in case 16, not only the stomach but also the aorta and heart are now on the right (Figure 18). These findings suggest complete situs inversus.<sup>71-73</sup>

**Case 17**

Case 17 simply shows that the heart is on the right (Figure 19); this is dextrocardia, as we have seen above, and is related to a looping anomaly during the embryologic development of the heart. A discrepancy between the position of the cardiac apex and that of the stomach is associated with a higher incidence of congenital heart defects (>75% in cases of visceral situs and >95% in cases of dextrocardia). This is in contrast to case 16 with complete situs inversus, in which both the stomach and the cardiac apex are on the right side. In situs inversus totalis, the likelihood of a cardiac anomaly is 2%.

**Figure 18.** Case 16. Four-chamber view showing a right-sided heart and the aorta (red) on the right side of the spine. The stomach (cyan) is also located on the right side. These findings represent complete situs inversus. This image is simplified because the level of the stomach and the 4-chamber view of the heart are depicted at the same level, whereas in reality the stomach is lower in the fetus.





**Figure 19.** Case 17. Four-chamber view showing dextrocardia. The aorta (red) and stomach (cyan) are located on the left.

## Conclusions

This short review highlights several of the anomalies that can be recognized on the 4-chamber view. This list is far from being exhaustive, and the drawings tend to simplify the findings, but these drawings could serve as a basis for those doing fetal echocardiography when they encounter an unusual finding.

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