Right and Left atrial isomerism. Two case reports

Abstract

The term isomerism refers to symetrical development of organs that are normally asymetrical. These characterize the heterotaxy syndromes. In heterotaxy syndromes the position of the organs are different from situs solitus. We present two cases of atrial isomerism which is a complex anomaly and rather difficult to diagnose in utero. For both cases, the death of the fetuses were achieved at 24 weeks of gestation. We concluded that survival of children with left atrial isomerism is higher than those with right isomerism. In this respect, the second case shows to become more appropiate for surgery.
Keywords:*isomerism, atrial situs, visceral situs, heterotaxy*

C. Ionescu[1], E. Bratilă[1], I. Pacu[1], M. Banacu[1], N. Bacalbașa[2]

1. Department of Obstetrics and Gynecology, "Carol Davila" UMF, Clinical Emergency Hospital Sf Pantelimon, Bucharest (Romania)
2. Assistant Prof. UMF "Carol Davila", Department of Obstetrics and Gynecology, "Carol Davila" UMF, "Cantacuzino" Hospital, Bucharest (Romania)

Correspondence: Dr. Cringu Ionescu e-mail: antoniuginec@yahoo.com

Introduction

In the ultrasound evaluation of the fetal heart the first step is establishing the fetal visceral situs. Determination of the atrial and ventricular situs is very important in clinical practice. Situs is the location or position that an organ occupies in a bilateral system of symetry[(1)]. In other words the term *situs* refers to the position of the heart and viscera relative to the midline. All normal or malformed hearts are made of 3 segments: atria, ventricles and great vessels. In the sequential segmental analysis it is mandatory that the cardiac segments are identified based on their anatomical characteristics.

Case reports

Case 1-a 22 year old primipara is refered for morphologic ultrasound at 22 weeks of gestation with no previous investigation with this pregnancy and she was diagnosed with left atrial isomerism. A second case (case 2) of a 41 year old secundipara is refered for morphologic ultrasound for a second opinion. This patients was already suspected with a right atrial isomerism.

In this respect, to determine the atrial situs it is necesary to establish the position of each atrium in relation to each other and it must be starting with the differentiation between left and right atria. The right and left atrial appendages are different, but are difficult to identifie with ultrasonography. The morphologically right atrium is characterized by a broad based triangular appendages and the left morphologically atrium has a narrow based, tubular, hooked left appendage.

There are three types of atrial situs and also three types of visceral situs. The three types of atrial situs are: 1. *situs solitus* or *usual atrial arrangement*; 2. *situs inversus* or *mirror image*; and 3. *situs ambiguous* that is left isomerism and right isomerism (in Greek language iso means "the same" and meros means "turn"). Also the three types of visceral situs are: 1. *situs solitus* that mean normal arrangement of vessels and organs in the body; 2. *situs inversus* that mean a mirror image of the vessels and organs in the body; 3. *situs ambiguous* (i.e. heterotaxic syndrome) that mean visceral and vessel malposition in the body (i.e. heterotaxy syndrome, in wheach heteros means different, and taxis means arrangement)[(2)].

In usual atrial arrangement or *situs solitus* there are the following: the morphologically right atrium is to the right of the left morphological atrium, inferior vena cava is to the right of the spine and more anterior than the aorta who is left sided, the stomach is on the left, the liver and portal vein is on the right.

In situs inversus there is an mirror image of the atria, vessels, organs, to *situs solitus* normal arrangements. Therefore, the morphologically right atrium is on the left side, inferior vena cava is on the left side and aorta is on the right, stomach is on the right side, and major hepatic lobe is on the left side[(3)].

In situs ambiguous there are two types of isomerism: right isomerism and left isomerism. In right isomerism there are two right morphologically right appendages, both sides of the body show the right morphology, contralateral position of the heart with stomach, and the abdominal vessels are on the same side of the spine (right or left). In left isomerism there are two morphologically left appendages on atria (Figure 1), both side of the body show the left morphology, con-

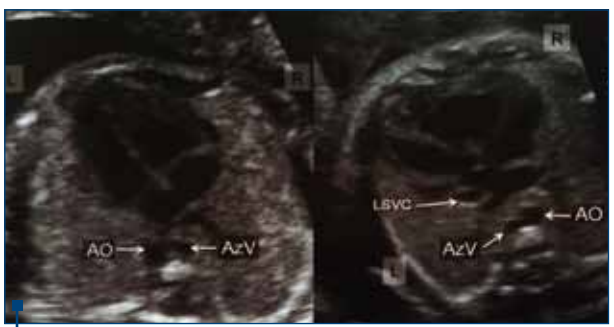

Figure 1. The 4th chamber view with left atrial isomerism and double vessel sign (LSVC=left superior vena cava, AzV=azygos vein, Ao=aorta)

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tralateral position of the heart with the stomach, the inferior vena cava is interrupted and the azygos vein is a structure posterior to the aorta and the two vessels are in front of the spine (in all four chamber view).

The current method of determination of fetal situs is the position of the stomach and heart, a careful attention should be adressed to the inferior vena cava and aorta below the diaphragm.

Establishing the fetal visceral situs and atrial situs

To deterime the fetal situs during the ultrasound examination we must fulfilled the following steps: Step 1. To determinate the presenting part. Step 2. To determinate the fetal lie within the uterus by obtaining a sagital view of the fetal spine. Step 3. To determinate the location of the fetal left side with regard to maternal abdomen, fetal left side is anterior, posterior, or right lateral or left lateral. Step 4. to have a transverse view of the fetal abdomen in which the stomach is on the left side, descending aorta is on the left and posterior, the inferior vena cava is anterior and to the right. Although determining that the stomach, descending aorta and cardiac apex are located on the fetal left side and the inferior vena cava located on the right side we can establish a normal situs visceral[4]. We can use also the method described by Brohnstein and contributors[2] which is called ‚right-hand', a rule for abdominal scanning and the left hand rule for transvaginal scanning. In these method, the palm of the hand corresponds to the face of the fetus and the examiner holds the hand according to the side of the fetal face, and the fetal heart and stomach are shown by the examiners thumb (Table 1).

There are two major situations in which we suspect a situs abnormality: in the 1st situation, the fetal heart and stomach are on opposite side of the body, and in the 2nd situation, a complex malformation of the heart is presented.

In both of our cases the first ultrasound sign was that the stomach and fetal heart were in opposite side.

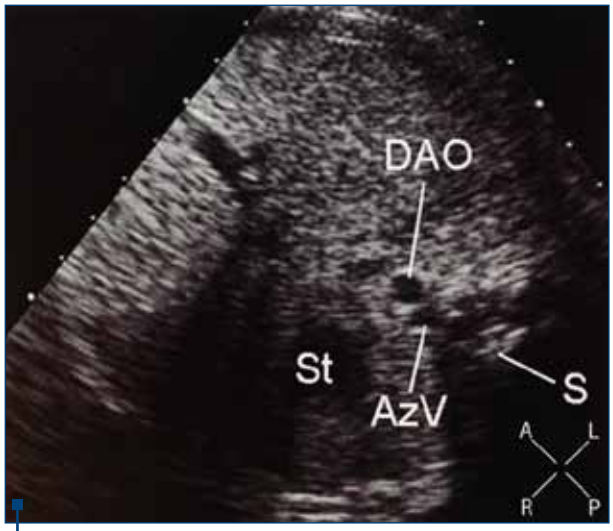

Figure 2. Left isomerism. Azygos vein is posterior to the aorta and stomach on the right (DAO=descending aorta, St= stomach, AzV= azygos vein, S= spine)

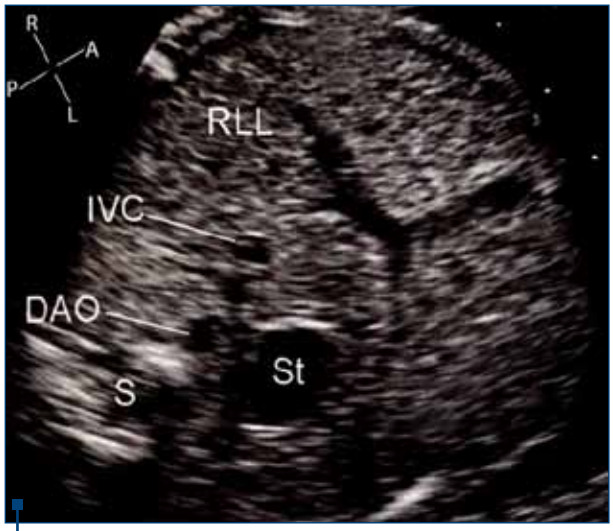

Figure 3. Right isomerism. The inferior vena cava and aorta are on the right side (RLL= right liver lobe, IVC= inferior vena cava, DAO= descending aorta, S= spine, St= stomach)

Table 1 The main types of visceral situs

Situs	Right side	Left side
Solitus	Morphologic right atrium Major hepatic lobe Inferior vena cava Trilobed lung Short bronchus	Morphologic left atrium Stomach Descending Aorta Bilobed lung Long bronchus
Inversus	Morphologic left atrium Stomach Descending aorta Bilobed lung	Morphologic right atrium Major hepatic lobe Inferior vena cava Trilobed lung
Ambiguous	Variable	Variable

Ultrasound diagnosis of left atrial isomerism (polisplenia)

In left atrial isomerism (case 1) there are double left sided structure with absence of right sided structure. Therefore, in left atrial isomerism we can have: shift of axis, absence of intrahepatic portion of inferior vena cava, complete heart block, left ventricular outflow obstruction.

In the situation of absence of intrahepatic portion of inferior vena cava (seen in 100% cases), the abdominal venous blood is drain in azygos system, and the azygos vein is dilated with its typical course side by side and posterior to the descending aorta[5,6], and not anterior as is the course in case of vena cava inferior present (Figure 2).

This condition is termed interruption of the inferior vena cava with azygos continuation and the main echographic feature is a double vessel sign in a axial cross section of the upper abdomen. Also this anomaly is evident in the four chamber view in which we can see behind the left atrium two vessels of similar size but different pulsatility. In a parasagital view of the abdomen and chest the azygos vein can also be seen posterior to the descending aorta. Having in the view the absence of right atrium with its sinus node sometimes this can create a complete heart block[7]. Other abnormalities seen in left atrial isomerism are: polysplenia (which was not diagnose on ultrasound), duodenal atresia or esophagian atresia, midline liver, absence of gallbladder.

Ultrasound diagnosis of right atrial isomerism (asplenia)

Right atrial isomerism (case 2) is associated with the presence of double right sided structure and the absence of left sided structure. So, in right atrial isomerism we have: shift of axis to the right, anomalous position of the abdominal aorta and the the inferior vena cava both positioned centrally or on the same side of the spine (Figure 3), unbalanced AVSD, malposition of great arteries, partial or total abnormal pulmonary venous connection.

For the diagnosis of both left or right atrial isomerism we can use the colour Doppler (Figure 4) and the 3D/4D dimensional ulltrasound.

Color Doppler can diagnose the vessel arrangement (Figure 4) and the cardiac malformation. That is the venous connections, great vessel and atrioventricular regurgitation. Also, it can diagnose ductus dependence.

Four dimensional echocardiography can be used in locating normal or abnormal veins. Using inversion mode with power Doppler or glass body mode we can see the interrupted inferior vena cava[8].

Differential diagnosis of isomerism must be made based on dextrocardia and *situs inversus*. In *situs inversus* the liver and inferior vena cava are on the left side of the fetus and the stomach and heart are on the right side. In dextrocardia the heart axis point to the right anterior thorax.

Discussion

The prognosis of left and right isomerism is poor depending of the severity of the anomaly detected, especially heart anomalies. Fetuses are at increased risk of dying in utero because of development of hydrops and of heart block. In cases of left isomerism with mild forms of cardiac malformation the prognosis is good. In the study of Takerazu[9], fetal left isomerism was associated with a mortality rate of 31% and only 3 out of 14 with right isomerism were survived. The isomerism is not associated with chromosomal anomalies and there is no need for karyotype method.

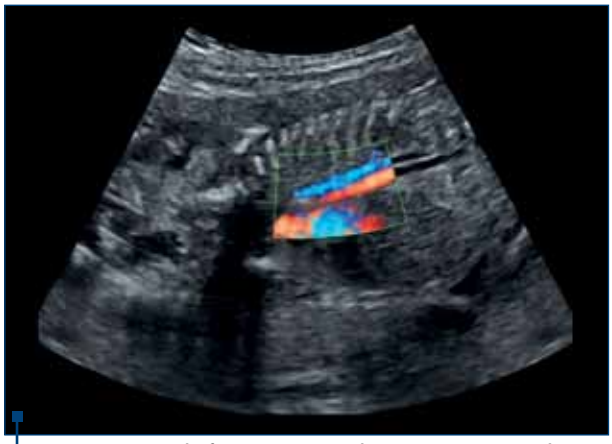

Figure 4. Interrupted inferior vena cava with azygos continuation (the reverse direction of blood flow in the azygos)

In both our cases in utero death was achieved at 24 weeks of gestation. With all this, survival of children with left atrial isomerism is higher than those with right isomerism. For the second case, the surgery is most reccomended, although only when total anomalous pulmonary venous connection appear, the indication is more clear. Sometimes, in the case of left atrial isomerism there is no need for a treatment.

Conclusions

In cases of biventricular correction the results are better than in cases of univentricular correctly surgery. The overall mortality in right and left atrial isomerism is 50%, but it depends of the type of heart malformation present. ■

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