Scientific letters

Echocardiographic features of fetal mesocardiac: a different heart

Características ecocardiográficas de la mesocardia fetal: un corazón diferente

To the Editor,

The position of the heart is systematically assessed during fetal cardiac examination. A change in heart position is uncommon and very important, as it may be associated with intracardiac and extracardiac structural anomalies.[1]

Mesocardia in an uncommon cardiac abnormality, in which the heart is positioned in the center of the thorax and its longitudinal axis lies in the thoracic mid-sagittal plane. The true incidence of this condition is unknown.

Mesocardia is usually associated with other structural cardiac abnormalities, but occasionally it is found alone. Most of the reported prenatal cases are related to heart defects or extracardiac anomalies. Displacement of the heart to the thoracic midline can occur secondary to an intrathoracic mass, pulmonary abnormalities, or diaphragmatic hernia.[1] Mesocardia has also been described in association with severe cardiac anomalies, heterotaxy, chromosome disorders, and genetic syndromes.[1,2]

Here, we describe 2 cases of mesocardia diagnosed prenatally in our prenatal diagnosis unit, together with their ultrasound characteristics.

The first case was the fifth pregnancy in a 33-year-old woman whose 4 previous pregnancies had culminated in 1 birth and 3 abortions. She came to our unit for early fetal echocardiography at 16 weeks' gestation. The fetal heart was in a midline position and showed an associated persistent left superior vena cava (LSVC). No additional cardiac or extracardiac structural abnormalities were detected. The heart showed an atypical morphology: The left ventricle had a rounded shape instead of the characteristic inverted cone, and the cardiac apex had an unusual appearance. The patient did not wish to undergo amniocentesis. There were no changes on fetal follow-up. At 36 + 6 weeks, a girl was born by natural vaginal delivery, with good clinical status and weighing 2230 g. Mesocardia and persistent LSVC were confirmed postnatally. At the time of writing, the child is 24 months of age and she has remained asymptomatic except for 1 episode of acute bronchiolitis.

The second case was the first pregnancy in a 30-year-old woman, who attended our prenatal diagnosis unit to undergo morphological ultrasound study. Mesocardia with an associated LSVC was observed. On careful anatomic examination, no other anomalies were noted. As in the previous case, the heart had an unusual morphology, showing a more rounded shape than normal. There were no additional findings on fetal follow-up, except for intrauterine growth retardation. At 41 + 2 weeks, a boy was born by forceps delivery, weighing 2450 g and with good clinical status. At the time of writing, he is 22 months old and has experienced an episode of acute bronchiolitis and several bouts of severe laryngitis requiring hospitalization.

In both these mesocardia cases, the heart showed an atypical morphology (figure 1). On postnatal echocardiography, there were no increases in cardiac trabeculation or myocardial changes in either infant.

Within the fetal thorax, a normal position of the heart and orientation of the interventricular septum with the midline are constant features. Variations in this pattern suggest the presence of a diaphragmatic, cardiac, or pulmonary anomaly. Assessment of the cardiac position in the fetal thorax is an important component of 4-chamber examination during routine imaging.[2]

Comparison of the morphology of hearts in mesocardia with those in levocardia shows a more rounded left ventricle than normal, without the typical inverted cone shape. The tip of the heart does not have the usual apical form, resulting in a morphology different from that of a heart in levocardia.

In 1971, Lev et al.[3] described the anatomic characteristics of midline hearts. These authors performed autopsy study of 13 cases and reported that the abnormal morphology has an embryologic origin. In normal conditions, after the heart has completely formed and reached a size of 20 to 25 mm, there is no true apex and it has a shape similar to that of a heart in mesocardia. In the following phase, the heart points toward the left or right and attains an apical shape (figure 2A). Lev et al. reported that the apex may be hidden or "absent" in midline hearts. In the area where the apex should be, there is a wide stretch of ventrally oriented muscle fibers. It is this distribution that confers the tear shape seen on chest radiology studies (figure 2B).

Allan et al.[4] reported that the cardiac apex points in an anterior direction at 9 weeks and rotates toward the fetal left at 11 weeks. McBrien et al.[5] evaluated the cardiac axis in 188 fetuses between 8 and 15 weeks of gestation and also concluded that the fetal cardiac axis has a midline position at 8 weeks' gestation and undergoes levorotation at the end of the first trimester. The normal, levorotated position is attained at approximately 12 weeks.

The looping that occurs in the fetal cardiac axis at the end of the first trimester is one of the last phases of cardiac embryogenesis. During normal looping, a complex series of changes takes place in the position of the heart.[6] At the beginning of this process, the ventricles have a craniocaudal relationship. In the final phase, the heart rotates in a counterclockwise direction around the basal-apical axis, with the right ventricle moving ventrally. This final phase and the relatively late changes occurring in the shape of the left ventricle can explain the change in the cardiac axis seen in early fetal echocardiography.[5] Thus, midline hearts that do not complete this final embryological phase show both a change in the cardiac axis and an abnormal apical morphology.

An associated persistent LSVC was present in both cases described, and 1 fetus showed intrauterine growth retardation. Persistence of the LSVC tends to be more common in hearts in mesocardia than those in levocardia associated with congenital heart disease.[3] Of the 13 cases described by Lev et al., 6 (46%) were associated with this vascular anomaly. Development of the innominate vein and LSVC involution occur at the end of cardiac morphogenesis, at the same time as looping of the cardiac axis.

https://doi.org/10.1016/j.rec.2019.08.011

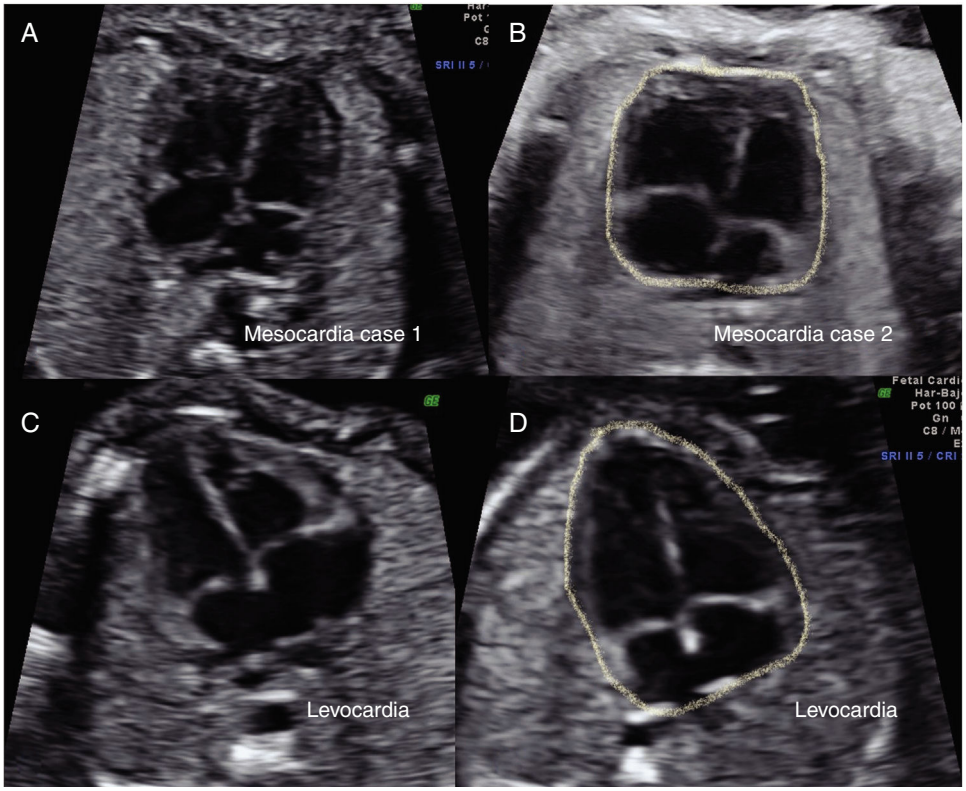

Figure 1. Comparison of heart morphology. Transverse view in 2 fetuses with mesocardia (A and B) and 2 fetuses with levocardia (C and D). In the fetuses with mesocardia (A and B), the heart is positioned at the center of the thorax and the interventricular septum is aligned with the thoracic midline. In both cases, the hearts have a squared-off shape that contrasts with the triangular shape of the hearts in levocardia (C and D).

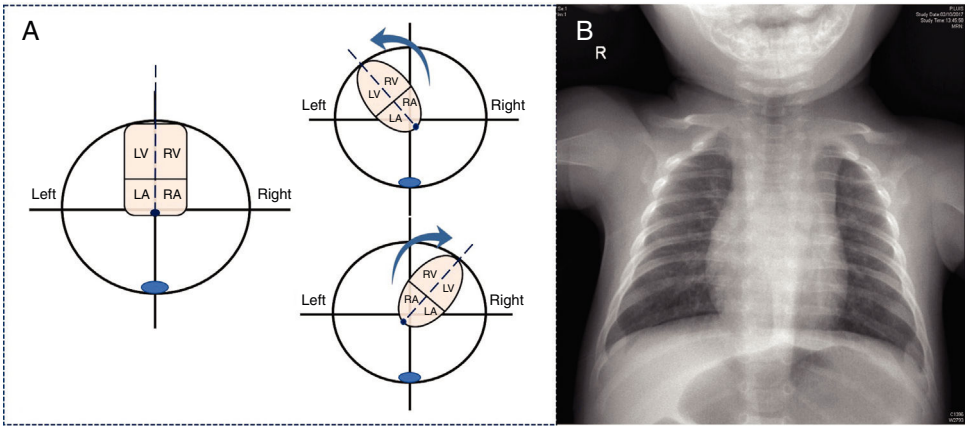

Figure 2. Morphology of the midline heart. A, diagram of the final phase of fetal embryology, when the cardiac axis changes and the heart acquires its final form. B, anteroposterior chest radiograph of the second case depicts the heart in the center of the thorax with a teardrop- or dewdrop-shaped silhouette. RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

Detection of primary mesocardia in the second trimester of gestation in a structurally normal heart is quite unusual. In most cases, mesocardia is associated with major cardiac or extracardiac defects; hence, meticulous study of the fetal anatomy should be performed. The 2 cases diagnosed in our unit showed the atypical morphology described by pathologists. A familiarity with the characteristics of these hearts facilitates prenatal treatment and parental counseling about this condition, and may avoid overdiagnosis of other cardiac conditions.

Raquel García Delgado,* Raquel García Rodríguez, Javier Segura González, María de Luis Alvarado, Margarita Medina Castellano, and José Ángel García Hernández

Servicio de Obstetricia y Ginecología, Hospital Universitario Materno Infantil de Canarias, Las Palmas de Gran Canaria, Spain

* Corresponding author:
E-mail address: rgdelgado1@hotmail.com (R. García Delgado).

Available online 16 November 2019

REFERENCES

1. Falkensammer CB, Ayres NA, Altman CA, et al. Fetal cardiac malposition: incidence and outcome of associated cardiac and extracardiac malformations. *Am J Perinatol.* 2008;25:277–281.
2. Allan LD, Lockhart S. Intrathoracic cardiac position in the fetus. *Ultrasound Obstet Gynecol.* 1993;3:93–96.
3. Lev M, Liberthson RR, Golden JG, Eckner FA, Arcilla RA. The pathologic anatomy of mesocardia. *Am J Cardiol.* 1971;28:428–435.
4. Allan LD, Santos R, Pexieder T. Anatomical and echocardiographic correlates of normal cardiac morphology in the late first trimester fetus. *Heart.* 1997;77:68–72.
5. McBrien A, Howley L, Yamamoto Y, et al. *Ultrasound Obstet Gynecol.* 2013;42:653–658.
6. Männer J. The anatomy of cardiac looping: a step towards the understanding of the morphogenesis of several forms of congenital cardiac malformations. *Clin Anat.* 2009;22:21–35.

https://doi.org/10.1016/j.rec.2019.08.011
1885-5857/

Catheter-based closure of residual leaks after percutaneous occlusion of the left atrial appendage with WATCHMAN device: two cases

Cierre percutáneo de fugas paravalvulares residuales después del cierre percutáneo de la orejuela izquierda con dispositivo WATCHMAN: serie de dos casos

To the Editor,

Left atrial appendage (LAA) closure with the WATCHMAN device (Boston Scientific; Minnesota, United States) is as effective as warfarin in preventing embolisms in patients with nonvalvular atrial fibrillation (NVAF) and is associated with a comparative reduction in bleeding events and cardiovascular mortality.[1] This procedure is a feasible alternative to oral anticoagulant (OAC) therapy for patients with a high bleeding risk.

Residual leakage around the device is an important limitation in LAA closure. The causes of this event include variability of the LAA ostium shape, underestimation of its size, device migration, and difficulty in covering several lobes.[2]

The optimal treatment for peridevice leakage following WATCHMAN implantation is uncertain. In patients who have undergone LAA surgical ligation, embolism rates are higher in those with residual communications than in those with complete closure.[3] It is accepted by consensus that leaks < 5 mm wide are not significant, and they are not considered a procedure failure.[4] Treatment for leaks > 5 mm in size is controversial. In contrast to the recommendation to maintain OAC indefinitely, some authors suggest percutaneous closure as a viable option, considering the patients' bleeding risk and the development of technical aids such as microwave use in transesophageal echocardiography (TEE).[5] Experience with this type of intervention is scarce and limited to small case series.[2] Here, we report on the first 2 cases in Spain of percutaneous closure of major peridevice leaks (> 5 mm) associated with WATCHMAN.

The first case was an 85-year-old man with permanent NVAF and indefinite OAC, a history of lower gastrointestinal tract bleeding, hypovolemic shock, and hemorrhagic stroke, and a CHA_2DS_2-VASc score of 6 and HAS-BLED score of 5. Implantation of a 24 mm WATCHMAN device was carried out, with adequate intraoperative compression and detection of an anterosuperior leak < 3 mm wide (figure 1A, arrow) that was treated conservatively. After 1 year with no incidents, the patient experienced an ischemic stroke in the left frontal territory. TEE showed proximal migration of the device and 2 leaks > 5 mm wide, one in an anterosuperior position (figures 1B,C, yellow arrow) and the other inferoposterior (figure 1B-D, white arrow). Possible causal mechanisms include poor initial fixation or posterior LAA dilation due to the initially incomplete closure, as occurs in cases of incomplete percutaneous closure. Because of the patient's contraindications for restarting OAC, percutaneous closure was decided. An 8.5-Fr Medium Curl Agilis sheath (Abbott; Illinois, United States) was placed in the left

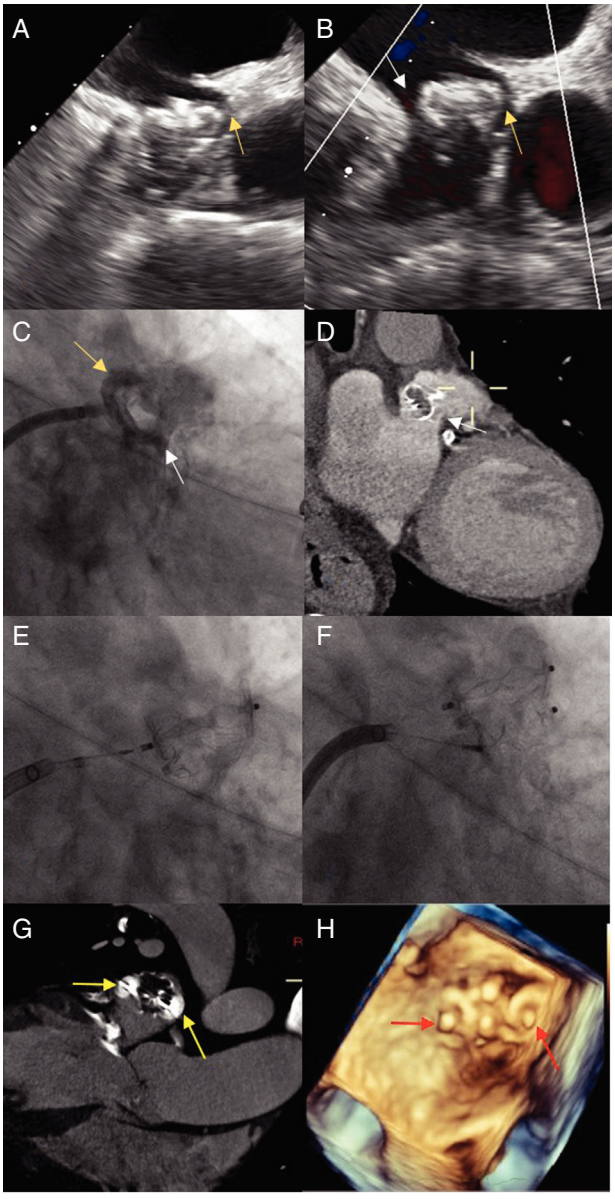

Figure 1. A: transesophageal echocardiography image showing an anterosuperior leak < 3 mm wide (yellow arrow) following the first procedure. B: transesophageal echocardiography image depicting an anterosuperior leak (yellow arrow) and inferoposterior leak (white arrow) after the patient experienced a stroke. C: fluoroscopy image showing the anterosuperior (yellow arrow) and inferoposterior (white arrow) paravalvular leak. D: computed tomography image of the inferoposterior leak (white arrow). E: fluoroscopy image depicting anterosuperior implantation of a 12-mm Amplatzer Vascular Plug II. F: fluoroscopy image showing posteroinferior implantation of a 12-mm Amplatzer Vascular Plug II. G: computed tomography image of the final position of the devices (yellow arrows). H: 3-dimensional ultrasound image of the final position of the devices (red arrows).