

An isolated fetal cor triatriatum dexter during a targeted anatomic survey at 22 weeks' gestation

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Summary

Cor triatriatum dexter is a rare cardiac malformation characterized by division of the right atrium into two compartments by a usually fenestrated membrane, whose degree of partitioning or septation is responsible for different clinical manifestations. Cor triatriatum dexter has been diagnosed incidentally in adults and children by echocardiography, surgery, or autopsy but has not previously been published in fetuses, apart from a report of two early-2nd-trimester fetuses aborted as a result of associated severe nuchal edema. To our knowledge, this is the first report of an isolated fetal cor triatriatum dexter that was diagnosed during ultrasound screening at 22 weeks of gestation.

KEY WORDS: cor triatriatum dexter, cor triatriatum dextrum, congenital heart disease.

Introduction

Cor triatriatum dexter is a rare congenital abnormality in which the right atrium is divided into two chambers by a membrane and is commonly associated with other right-sided cardiac abnormalities. It occurs in 0.1-0.4% of all patients with congenital heart disease (1-3). In most cases, cor triatriatum is recorded at necropsy or is an incidental finding at surgery, transthoracic, or transesophageal echocardiography study (4).

The first report of this entity was a postmortem description by Church in 1868 (5).

Maroun et al. firstly described a report of two cases of

fetal cor triatriatum dexter associated with nuchal edema in early second trimester (6).

We present the first case of a fetal isolated cor triatriatum dexter that was diagnosed during ultrasound screening at 22 weeks of gestation.

Case report

A 31 year-old gravida 1, para 0, woman presented to the Prenatal Diagnosis Center, Artemisia in Rome to perform a screening ultrasound at 22 weeks of gestation. Neither maternal nor fetal pathologies were found during the entire pregnancy. The patient's past medical history was entirely noncontributory, with no cardiac risk factors or any previous medical/surgical intervention. Family history was reportedly negative for cardiac disease.

The sonographic examination of the fetal cardiac morphology revealed the presence of a membrane stretched between the medial and lateral walls of right atrium in the apical four-chamber view (Figure 1).

The fetal heart was evaluated using the sequential segmental approach that included:

Determination of visceral situs and position of the heart;
Identification of the morphologies of each component of each segment;

Assessment of the spatial relationships between the component of each segment:

- Atrial relationship or situs
- Ventricular relationship
- Great arterial relationship

Determination of the connections between the segment at the atrio-ventricular and ventriculo-arterial junctions:

- Atrio-ventricular connection
- Ventriculo-arterial connection

Evaluation of the associated malformation at each cardiac segment.



Figure 1

A Pulsed Wave Color Doppler technique was also used to assess the cardiac flows.

The careful study of cardiac morphology did not reveal further complex congenital anomaly.

Fetal morphology and biometric features were normal and appropriate for gestational age.

All previous fetal ultrasound examinations were normal, as well as the invasive prenatal diagnosis by amniocentesis, that revealed a normal female karyotype (46,XX).

Discussion

Cor triatriatum dexter is a rare cardiac malformation characterized by division of the right atrium into two compartments by a usually fenestrated membrane representing the persistence of remnants of the right valve of the embryonic sinus venosus (7).

During embryogenesis, the right horn of the sinus venosus gradually gets incorporated into the right atrium to form the smooth posterior portion of the right atrium whereas the original embryologic right atrium forms the trabeculated anterior portion. The connection between the right horn of the sinus venosus and the embryologic right atrium is the sinoatrial orifice which has two valvular folds on each side, the right and left venous valves (8). At some point during this incorporation, the right valve of the right horn of the sinus venosus divides the right atrium into two. This right valve forms a membrane that serves to direct the oxygenated venous return from the inferior vena cava across the foramen ovale to the left side of the heart during fetal life (9). Normally, the valve regresses by approximately 12 weeks of gestation: its cephalic portion develops into the crista terminalis and the caudal portion is divided into the valve of the inferior vena cava (the eustachian valve) and the valve of the coronary sinus (the thebesian valve) (10). With an incomplete regression, a fenestrated or an unfenestrated membrane may persist in the right atrium.

Abnormal persistence of the right sinus venosus valve is classified into two main categories: those with web-like remnants of the right valve (also known as Chiari net) and those with a partition by a membrane between the venous and trabeculated portions of the right atrium, that is, cor triatriatum dexter (11, 12).

In a review by Doucette and Knoblich, the most common location of this membrane was to the right of the superior vena cava, coronary sinus, and inferior vena cava and the second most common pattern found was when the membrane was to the left of the coronary sinus but to the right of the other two venous vessels (13).

Cor triatriatum dexter has varying clinical manifestations depending on the degree of partitioning or septation of the right atrium. When the septation is mild, the condition is often asymptomatic and is an incidental finding frequently made at postmortem examination; more severe septation can cause right-sided heart failure and elevated central venous pressures due to obstruction of the tricuspid valve, the right ventricular outflow tract, or the inferior vena cava. Presumably the membrane fa-

vors maintenance of blood flow from the inferior vena cava in the direction of the interatrial septum and obstructs the flow to the right ventricle.

Conclusion

Cor triatriatum dexter has been diagnosed in adults and children by echocardiography, surgery, or autopsy but has not previously been published in fetuses, apart from a report of two early-2nd-trimester fetuses aborted as a result of associated severe nuchal edema.

To our knowledge, this is the first report of an isolated fetal cor triatriatum dexter that was diagnosed during ultrasound screening at 22 weeks of gestation.

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