Images in Cardiovascular Medicine

Clear Morphological Depiction of Cor Triatriatum in a Child by Transthoracic 3-Dimensional Echocardiography

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A 3-year-old boy had a history of cor triatriatum, which was diagnosed incidentally when he was 1 year old during workups for failure to thrive and funnel chest. Initial echocardiographic evaluation revealed no evidence of obstruction through the orifice (mean pressure gradient, 3 mm Hg). However, progressive flow obstruction was noted at follow-up. At 3 years of age, 2-dimensional echocardiography at the apical 4-chamber view demonstrated a membrane-like structure traversing the left atrium (LA) (Figure 1A; see Movie I in the online-only Data Supplement). Doppler echocardiography revealed a mean pressure gradient of 8.9 mm Hg across the orifice of this membrane (Figure 1B). However, depiction of the relationships between this membrane and the surrounding structures was not clear. Therefore, trans thoracic 3-dimensional echocardiography (3DE) was performed (iE33; Philips, Andover, MA) with an X-7 matrix array transducer. The 3DE images cropped from the posterior and superior aspects of the LA toward the atrioventricular groove clearly showed that this membrane extended from the fossa ovalis area of the atrial septum to the LA free wall (Figure 2A; see Movie II in the online-only Data Supplement). Therefore, the LA was divided into 2 chambers (Figure 2B; see Movie III in the online-only Data Supplement). The communication between these 2 chambers was through a single, relatively small, horseshoe-shaped orifice, which was located near the atrial septum (Figure 2A; see Movie II in the online-only Data Supplement). Pulmonary veins connected to the posterior superior chamber (Figure 2A). In addition, a small secundum-type atrial septal defect was found within the anterior inferior chamber, which connected to the mitral valve (Figure 2C). Color 3DE further confirmed the location and size of this orifice, which was $\approx 0.43 \text{ cm}^2$ in area (Figure 2D; see Movie IV in the online-only Data Supplement). Multidetector computed tomography showed similar findings (Figure 3A and 3B). Cardiac catheterization revealed a pressure gradient of 12 mm Hg across the cor triatriatum (pulmonary artery wedge pressure, 15 mm Hg; mean left atrial pressure, 3 mm Hg). The patient underwent surgery for excision of cor triatriatum, repair of the atrial septal defect, tricuspid valve plasty, and correction of the funnel chest 4 months later. He had an uneventful perioperative course. Postoperative echocardiography showed complete excision of the membrane.

Cor triatriatum is a rare congenital cardiac anomaly accounting for 0.1% to 0.4% of all cardiac defects. The LA is divided into 2 chambers by an abnormal fibromuscular septum, which probably results from failure of resorption of the common pulmonary vein. Typically, the pulmonary veins drain into the posterior superior chamber, and the anterior inferior chamber drains into the left ventricle via the mitral valve. The 2 chambers are connected through 2 orifices in the septum. Clinical symptoms are related to the degree of restriction of blood flow, which depends on the size and number of fenestrations on the membrane. Surgical resection of the membrane is the treatment of choice for patients with significant flow obstruction. Theoretically, transesophageal echocardiography would provide better visualization of the cor triatriatum and its orifice. However, the invasiveness and long procedure time are major drawbacks of transesophageal echocardiography, especially for children. In this case, we demonstrated that transthoracic 3DE could provide accurate diagnosis of cor triatriatum by detailed appreciation of its relation with the atrial septum and the pulmonary veins, as well as the location and size of the orifice. These posterior cardiac structures could be visualized clearly by 3DE in children, whose echocardiographic image quality was usually superior to that in adults. Furthermore, experience from this pediatric case highlights the potential value of transthoracic 3DE in providing better appreciation of the anatomy and nearby structures around the cor triatriatum compared with the conventional echocardiography.

Disclosures

None.

References


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The online-only Data Supplement is available with this article at http://circ.ahajournals.orglookup/suppl?doi:10.1161/CIRCULATIONAHA.113.004075/DCl.

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(Circulation. 2013;128:e395-e396.)

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Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.113.004075


Figure 1. A, Transthoracic 2-dimensional echocardiography from the apical 4-chamber view shows a membrane-like structure within the left atrium, dividing it into posterior–superior (PS) and anterior–inferior (AI) chambers. B, Color Doppler echocardiography reveals significant flow acceleration across the membrane (estimated mean pressure gradient, 8.9 mm Hg).

Figure 2. A, Transthoracic 3-dimensional echocardiography (3DE) image viewed from the posterior and superior aspect of the left atrium (LA) toward the atrioventricular groove shows that a membrane (arrowheads) extended from the fossa ovalis area of the atrial septum to the LA free wall. Pulmonary veins are connected to the posterior–superior chamber. There is a single, horseshoe-shaped orifice (arrow) on the membrane near the atrial septum. B, 3DE image viewed from lateral aspect of the LA clearly shows that the LA is divided into posterior–superior and anterior–inferior chambers by this membrane. C, A small secundum-type atrial septal defect (asterisk) is found within the anterior–inferior chamber. D, Color 3DE image shows that the communicating orifice on the membrane is ~0.43 cm² in area (arrow). AO indicates aorta; AI, anterior–inferior chamber; LV, left ventricle; PS, posterior superior chamber; and PV, pulmonary vein.

Figure 3. A, Multidetector computed tomography demonstrates a membrane (arrowheads) dividing the LA into 2 chambers. B, Pulmonary veins drain into the posterior–superior chamber. AI indicates anterior–inferior chamber; PS, posterior superior chamber; and PV, pulmonary vein.
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*Circulation*. 2013;128:e395-e396
doi: 10.1161/CIRCULATIONAHA.113.004075
*Circulation* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/128/20/e395

Data Supplement (unedited) at:
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**Movie Legend**

**Movie 1.** Transthoracic 2-dimensional image from apical 4-chamber view shows a membrane extending from fossa ovalis area of the atrial septum to LA lateral wall, dividing the LA into 2 chambers. A small atrial septal defect is found between the anterior inferior chamber and the right atrium. Best viewed with Windows Media Player.

**Movie 2.** Transthoracic 3DE image viewed from the posterior and superior aspect of the LA toward the atrioventricular groove demonstrates a single horseshoe-shaped orifice on the membrane. Best viewed with Windows Media Player.

**Movie 3.** Transthoracic swivel 3DE image clearly depicts the spatial relationship between this membrane and the surrounding structures, as well as the location of communicating orifice. Best viewed with Windows Media Player.

**Movie 4.** Transthoracic multiplane color 3DE image demonstrates the flow across this membrane, marking the exact location and size of the orifice. Best viewed with Windows Media Player.