International Journal of Cardiology 166 (2013) e43–e47

Contents lists available at SciVerse ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Letter to the Editor

Three-dimensional echocardiography for cor triatriatum in adults ☆

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ARTICLE INFO

Article history:
Received 13 December 2012
Accepted 18 January 2013
Available online 22 February 2013

Keywords:
3-Dimensional echocardiography
Cor triatriatum
Adults

Cor triatriatum is a very rare congenital abnormality. In this malformation the left atrium (LA) is divided by a fibromuscular membrane into an upper (supero-posterior) chamber containing the pulmonary veins and their confluence and a lower (infero-anterior) chamber housing the true LA and left atrial appendage. The two chambers generally communicate through one or more openings in the intra-atrial membrane. The age at presentation and clinical symptoms is related to the degree of pulmonary venous obstruction. In the majority of cases it is diagnosed in neonatal period or early infancy, whereas adult cases are very rare and tend to be asymptomatic because the dividing membranes are often non-obstructive. Atrial septal defect or patent foramen ovale is present in 70–80% of patients with cor triatriatum [1–3]. Three-dimensional echocardiography (3DE) provides additional information such as the morphology, the size, and the number of openings of the dividing membrane and its spatial relationship to pulmonary veins, left atrial appendage and inter-atrial septum. We report 3 cases of non-obstructive cor triatriatum in adults whereby 3DE provides incremental information about the membrane, its relation to adjacent structures and associated cardiac anomalies.

Patient 1. A 67-year-old woman with a history of hypertension, ventricular septal defect (VSD) repaired in childhood and permanent pacemaker implantation for bradycadia received echocardiography because of new-onset palpitation (Fig. 1). Apical 4-chamber view on transthoracic echocardiography (TTE) showed a membrane-like structure (arrows) in the LA but the image was suboptimal (1A). Transoesophageal echocardiography (TEE) clearly showed a fibromuscular membrane (arrows) partially dividing the LA into 2 chambers on the mid-esophageal long-axis view (1B). There was no turbulent flow on color Doppler (1C) to suggest obstruction. When viewed from the atrial ('surgical') perspective on 3D TEE, the membrane (arrows) extended from the lateral side of the LA, partially dividing the LA into 2 chambers (1D). The upper chamber receives the left pulmonary vein and the lower chamber contains the left atrial appendage (1E). The mitral valve has normal structure and function. Her left ventricular function was normal and there was no residual VSD shunt. Patent foramen ovale (PFO), atrial septal defect (ASD) and anomalous venous connections were ruled out and she was incidentally found to have non-obstructive cor triatriatum. LV = left ventricle; RV = right ventricle; RA = right atrium; Ao = Aorta; AV = aortic valve; MV = mitral valve; LUPV = left upper pulmonary vein; LAA = left atrial appendage.

Patient 2. Fig. 2 showed the echocardiographic images of a patient with cor triatriatum and patent foramen ovale. She is a 27-year-old previously fit and healthy woman who received echocardiography because of a heart murmur detected during her pregnancy. Apical 4-chamber view on TTE showed a membrane-like structure (arrows) in the left atrium (2A). Color Doppler showed no evidence of turbulent flow suggestive of obstruction (2B). TEE clearly revealed a fibromuscular membrane (arrows) dividing the left atrium into 2 chambers on the mid-esophageal 5-chamber view (2C) but on the mid-esophageal commissural view (2D), the membrane (arrows) only partially divided the left atrium into 2 chambers. On 3D TEE, the membrane (arrows) could be clearly visualized separating the left atrium into 2 chambers where the upper chamber received the left upper pulmonary vein and the lower chamber contained the left atrial appendage (2E & 2F). The mitral valve had normal structure and function. Biventricular functions were also normal. There was a small patent foramen ovale (PFO) with a left to right shunt on color Doppler (2G). The PFO was located superior to the membrane (arrows). Atrial septal defect and anomalous venous connections were excluded. The patient did not undergo further intervention as she remained asymptomatic. LV = left ventricle, LA = left atrium; RV = right ventricle; RA = right atrium; Ao = aorta; LAA = left atrial appendage; LUPV = left upper pulmonary vein; AV = aortic valve; IAS = inter-atrial septum; RVOT = right ventricular outflow tract.

Patient 3. Fig. 3 showed the echocardiographic images of a patient with cor triatriatum and right ventricular volume overload secondary to a large patent foramen ovale (Fig. 3). He is a 47-year-old man with a history of hypertension presented with increasing shortness of breath. 2D TTE demonstrated right atrial and ventricular enlargement and elevation of pulmonary arterial systolic pressure to 45 mm Hg. TEE showed a membrane-like structure (arrows) in the left atrium consistent with the diagnosis of non-obstructive cor triatriatum (3A). Color Doppler revealed a large left-to-right shunt from the upper left atrial chamber to right atrium through a patent foramen ovale (PFO) (3B). Transcatheter closure of the PFO was indicated in view of the enlarged right heart and pulmonary hypertension. However, repeated attempts to deploy the left disk of the 25 mm Amplatzer PFO occluder

☆ Disclosure: None.

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http://dx.doi.org/10.1016/j.ijcard.2013.01.181

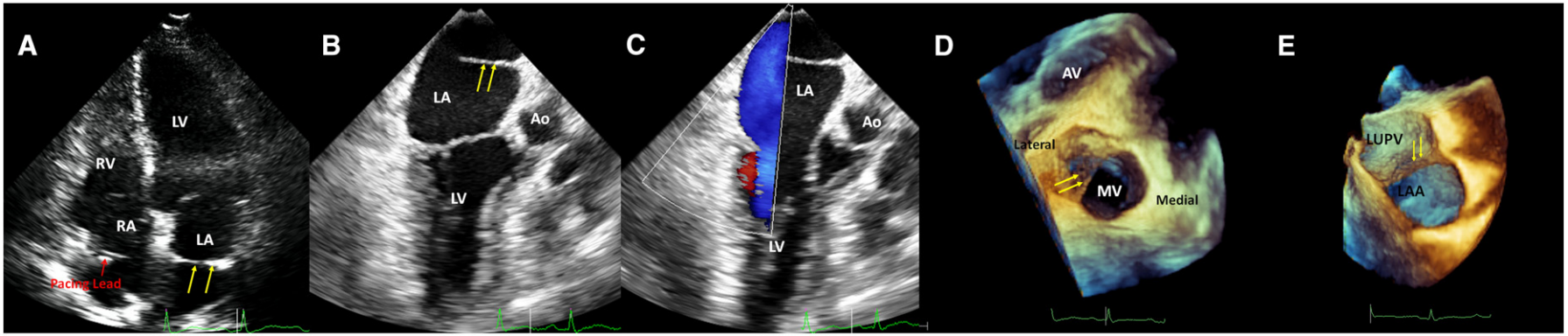

Fig. 1. Echocardiographic images of a patient with asymptomatic cor triatriatum.