

## Brief Report

# Usefulness of real-time three-dimensional trans-oesophageal echocardiography for detection of isolated unroofed coronary sinus

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**Abstract** We report a case of unroofed coronary sinus not associated with the persistent left superior vena cava. Definite diagnosis of the unroofed coronary sinus was obtained by trans-oesophageal echocardiography, which revealed the unroofed portion with left-to-right shunt. Real-time three-dimensional trans-oesophageal echocardiography could show the whole pictures of the defect, which was useful information for surgical repair.

**Keywords:** Unroofed coronary sinus; three-dimensional echocardiography; trans-oesophageal echocardiography

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## Case report

A 34-year-old woman was pointed out to have mild elevated right ventricular pressure by transthoracic echocardiography during her pregnancy at the general hospital. After 1 week of delivery, she had shortness of breath, and was admitted to the hospital with a diagnosis of pulmonary hypertension. Right ventricular pressure was estimated to be 70 mmHg. Her symptom improved with medical therapy, mainly with diuretics. She was diagnosed as having primum atrial septal defect by transthoracic echocardiography. Cardiac catheterisation revealed step-up of oxygen saturation at the right atrium with slight elevation of pulmonary artery pressure of 35/5(21) mmHg. The calculated pulmonary-to-systemic blood flow ratio was 2.7 and the pulmonary vascular resistance was 1.3 Woods unit m<sup>2</sup>.

She was referred to our hospital for the purpose of surgical repair. Transthoracic echocardiography at our hospital revealed dilation of the right atrium, right ventricle, and coronary sinus, although the atrial septum was intact. Contrast echocardiography with injection of contrast medium through the left

arm showed a negative jet from the coronary sinus towards the right atrium, indicating no association of persistent left superior vena cava.

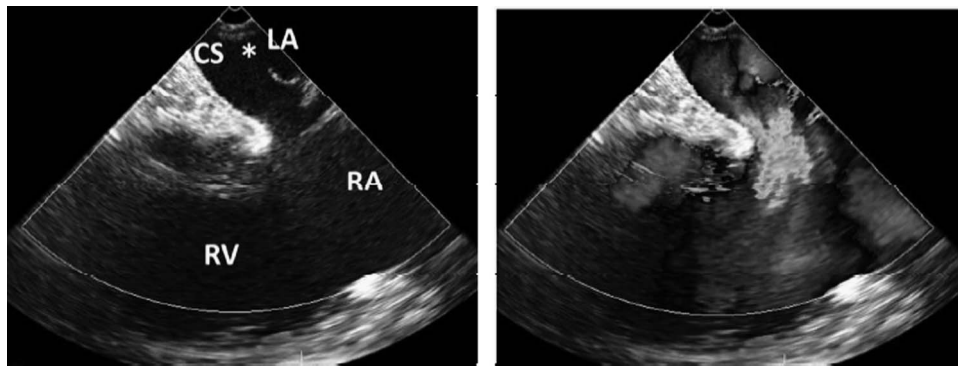
Owing to the fact that we suspected the presence of unroofed coronary sinus, trans-oesophageal echocardiography was performed. It clearly showed the defect between the left atrial wall and dilated coronary sinus with a shunt from the left atrium to coronary sinus using colour Doppler technique (Fig 1). Real-time three-dimensional trans-oesophageal echocardiography clearly demonstrated the whole picture of the unroofed portion with a defect size of 17 × 15 mm (Fig 2). Cardiac computed tomography also showed a communication between the left atrium and coronary sinus. Computed tomography also confirmed normal pulmonary venous return and absence of persistent left superior vena cava.

Closure of the defect of unroofed coronary sinus was performed using cardiopulmonary bypass with cardiac arrest. Through the incision of the right atrium, coronary sinus was cut back to the defect, and the whole defect was closed using autologous pericardial patch. The post-operative course was uneventful.

## Discussion

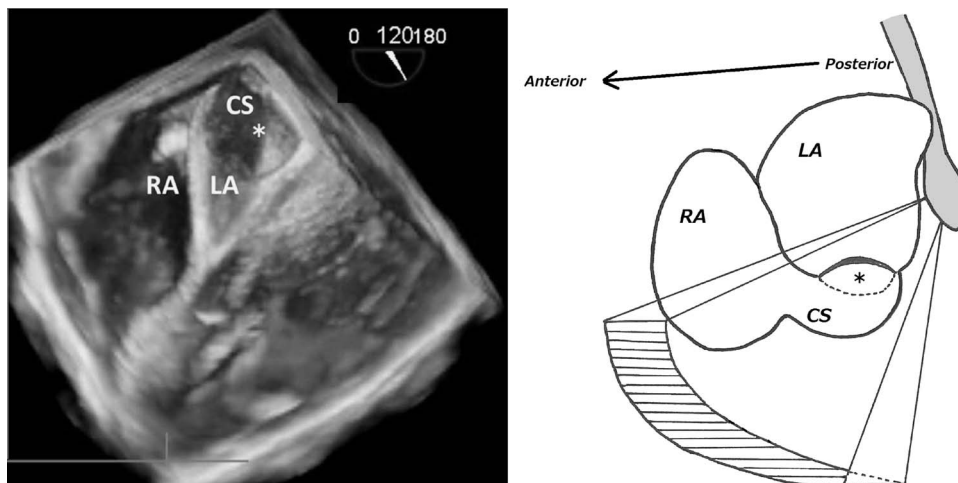
Unroofed coronary sinus is a very rare congenital heart defect, in which a communication exists

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**Figure 1.**

*Transesophageal echocardiography demonstrates the communication between left atrium and dilated coronary sinus. Color Doppler imaging reveals shunt flow from left atrium to right atrium through the unroofed coronary sinus. LA = left atrium; CS = coronary sinus; RA = right atrium; RV = right ventricle.*



**Figure 2.**

*Real-time three-dimensional echocardiography shows overview of unroofed portion from left atrium side. Right supplemental figure give an explanation the positional relationship between transesophageal echocardiography probe and unroofed coronary sinus with a sagittal view. LA = left atrium; CS = coronary sinus; RA = right atrium*

between the left atrium and coronary sinus. It is frequently associated with persistent left superior vena cava (75%). Its morphology has been classified into four groups – type I: completely unroofed with persistent superior vena cava; type II: completely unroofed without persistent left superior vena cava; type III: partially unroofed mid-portion; type IV: partially unroofed terminal portion.<sup>1</sup>

The clinical manifestation of this disease includes heart failure caused by chronic left-to-right shunt and increased risk of paradoxical cerebral infarction, although the clinical signs and symptoms are generally non-specific.

Transthoracic echocardiography is the most widely used imaging modality for the diagnosis of unroofed coronary sinus. Dilatation of coronary sinus with right atrium and right ventricle volume overload is a suggestive finding for unroofed coronary sinus when

we cannot find atrial septal defect, but it is often difficult to visualise the unroofed portion in spite of the use of colour Doppler technique. Multi-detector row computed tomography and cardiac magnetic resonance imaging may be indispensable imaging tools because they afford excellent anatomical information near the defect and combined lesions,<sup>2</sup> although its application for diagnosis of cardiac anomalies is still not popular.

Trans-oesophageal echocardiography is considered to be a very useful modality for the observation of posterior aspect of cardiac structures, such as coronary sinus and left atrium. It is capable of detecting precise unroofed portion and its anatomical relationship with the orifice of coronary sinus.<sup>3</sup> According to the development of technology, real-time three-dimensional trans-oesophageal echocardiography has been getting popular, especially for pre-operative

and intra-operative analysis of mitral valve morphology for mitral valve repair surgery.<sup>4</sup> In the field of congenital heart disease,<sup>5</sup> three-dimensional echocardiography has been widely used in some situations such as catheter intervention of atrial septal defect occlusion.<sup>6</sup>

Recently, the usefulness of real-time three-dimensional transthoracic echocardiography and trans-oesophageal echocardiography for unroofed coronary sinus has been reported as case reports.<sup>7,8</sup> In our case, real-time three-dimensional trans-oesophageal echocardiography clearly revealed the unroofed portion of the coronary sinus, which is an accurate diagnosis of type III unroofed coronary sinus without persistent left superior vena cava. Successful and uneventful surgical repair could be performed according to the precise diagnosis associated with its morphology.

We believe that real-time three-dimensional echocardiography might be a useful and powerful modality for the precise diagnosis of isolated unroofed coronary sinus.

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### Conflicts of Interest

None.

### Supplementary materials

To view supplementary material for this article, please visit <http://dx.doi.org/10.1017/S1047951113002400>.

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