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Partial Atrioventricular Septal Defect Assessed by Real-Time Three-Dimensional Echocardiography: A Case Report

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Abstract

A 48-year-old woman was admitted with a heart murmur and increased difficulty in breathing. Two-dimensional echocardiography revealed a defect in the lower part of the atrial septum[(primum atrial septal defect(ASD)]and a pouch at the interventricular septum. Color Doppler echocardiography detected grade 3/4 mitral regurgitation. Real-time three-dimensional echocardiography(RT-3DE)revealed a cleft in the anterior leaflet of the mitral valve toward the tricuspid valve, and the ASD located near the atrioventricular valves with 14 mm in minor axis. Color Doppler three-dimensional echocardiography disclosed left-to-right ASD shunt toward the atrial posterior wall. No shunt through the pouch at the membranous part, left ventricular outflow obstruction, or partial anomalous pulmonary venous connection was observed. RT-3DE is quite useful to describe complicated congenital heart disease.

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Key Words

■**Echocardiography, transthoracic**(**three-dimensional**)　■**Septal defects**(**atrioventricular**)

INTRODUCTION

Atrioventricular septal defect(AVSD), which is also called the endocardial cushion defect, involves the incomplete development of the inner tissues of the heart. Adult patients over 40 years old with AVSD have low risk in surgical repair and long-term survival is excellent,[1] although age at operation, preoperative mitral valve function, presence of atrial fibrillation and moderately elevated pulmonary vascular resistance are independent prognostic factors of surgical repair.[2] Understanding of the morphological type is important for the management of mitral regurgitation.[3]

The recent development of noninvasive imaging technology finally allows real-time three-dimensional volume rendering with transthoracic echocardiography. Because of the advantages, real-time three-dimensional echocardiography(RT-3DE)has been used in observing the complicated anatomy of congenital heart disease.[4,5] We report a case of an aged patient with partial AVSD examined with RT-3DE.

CASE REPORT

A 48-year-old woman, who had a heart murmur,

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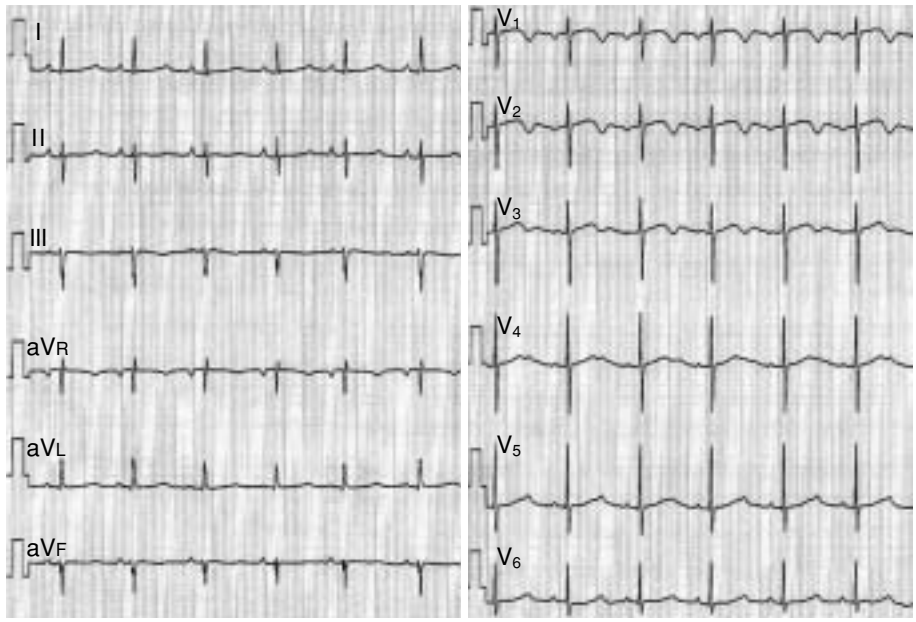

Fig. 1 Electrocardiogram

was admitted to our hospital because of increased difficulty in breathing. She had no remarkable medical history and no family history of cardiovascular disease. Her blood pressure was 110/60 mmHg, and heart rate was 82 beats/min regular. A grade 3/6 systolic ejection murmur at the second left sternal border, and fixed splitting of the S_2 with accentuated P_2 and S_3 increased in inspiration were detected. No rales were auscultated in the lung fields. Twelve-lead electrocardiography showed sinus rhythm, left axis deviation, left atrial overload (**Fig. 1**). Chest radiography showed bilateral pulmonary artery dilation and cardiothoracic ratio of 54% (**Fig. 2**).

Two-dimensional echocardiography revealed preserved left ventricular contractility (left ventricular end-diastolic diameter, end-systolic diameter and fractional shortening were 41, 21 mm and 49%, respectively). The enlarged left atrium and right heart were observed. A defect was present in the lower part of the atrial septum. Pouch formation was noticed at the superior end of interventricular septum. Color Doppler echocardiography showed grade 3/4 mitral regurgitation jet.

RT-3DE (Sonos 7500, Philips) was performed using the matrix probe (2−4 MHz). A full volume was acquired from the apical view for three-dimensional reconstruction. The three-dimensional images showed the cleft in the anterior leaflet of the mitral valve toward the tricuspid valve (**Fig. 3**). The atrial septal defect (ASD) located near the atrioventricular valves was 14 mm in diameter (**Fig. 4**). The left-to-right ASD shunt toward atrial posterior wall was viewed with color Doppler three-dimensional echocardiography (**Fig. 5**). The pouch formation was noticed at the superior end of interventricular septum. Neither left ventricular outflow obstruction nor partial anomalous pulmonary

Fig. 2 Chest radiograph

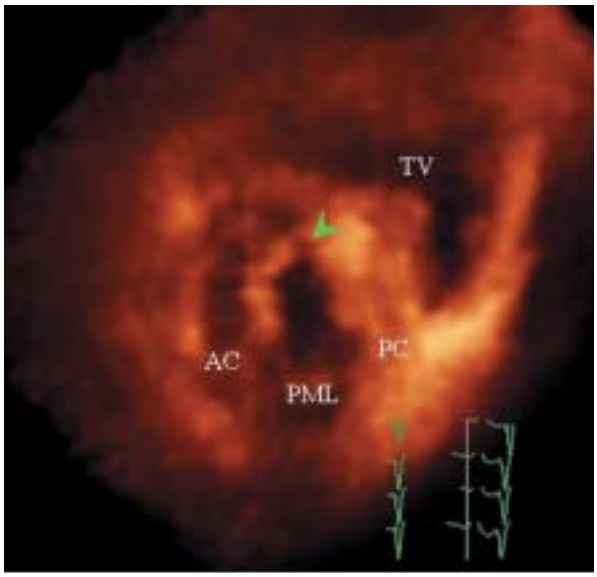

Fig. 3 Three-dimensional echocardiogram
The cleft (*green arrowhead*) was present in the anterior leaflet of the mitral valve.
TV = tricuspid valve; AC = anterolateral commissure of mitral valve; PML = posterior leaflet of mitral valve; PC = posteromedial commissure of mitral valve.

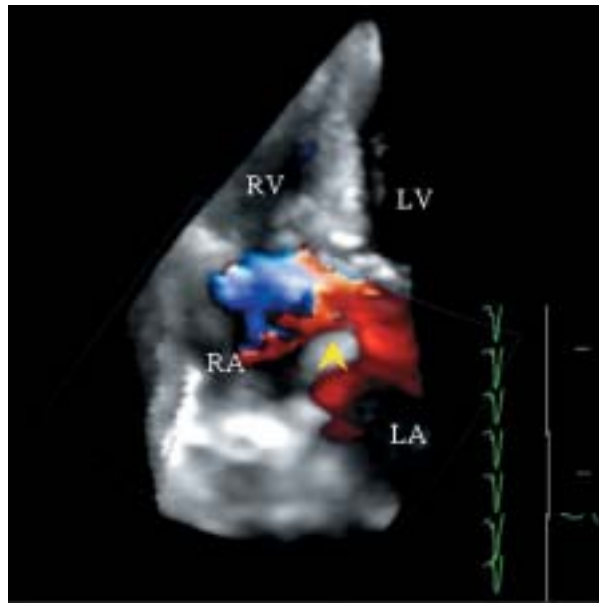

Fig. 5 Color Doppler three-dimensional echocardiogram
The left-to-right atrial septal defect (*yellow arrowhead*) shunt is seen toward the atrial posterior wall.
Abbreviations as in Fig. 4.

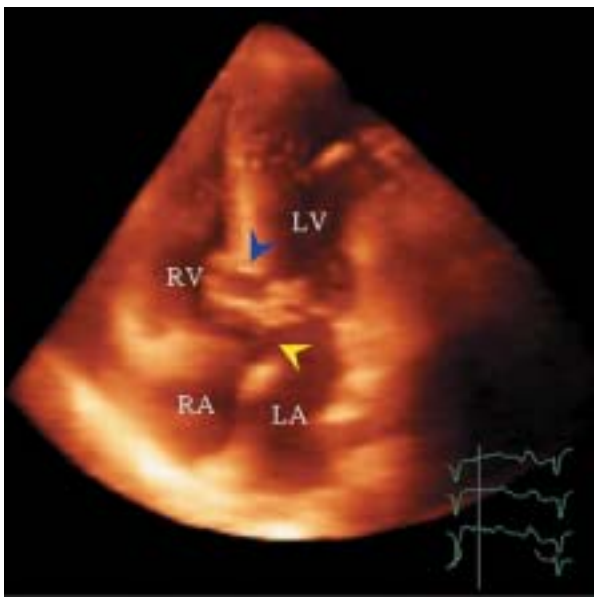

Fig. 4 Three-dimensional echocardiogram
Atrial septal defect (*yellow arrowhead*) is present near the atrioventricular valves and a pouch formation (*blue arrowhead*) is identified at the interventricular septum.
RV = right ventricle; LV = left ventricle; RA = right atrium; LA = left atrium.

venous connection was observed.

Right heart catheterization demonstrated pulmonary arteriolar resistance of 2.6 Wood's units, O_2 step up in the right atrium, and pulmonary to systemic flow ratio (Qp/Qs) of 3.01. Coronary angiography showed no significant stenosis. Left ventriculography showed normal left ventricular wall motion (ejection fraction = 54%), grade Ⅱ mitral regurgitation, and typical goose-neck sign.

Accordingly, the diagnosis was partial AVSD. She was indicated for surgical repair of the mitral valve cleft and primum ASD according to the surgical indication of ASD.

DISCUSSION

Partial AVSD is a form of AVSD characterized by two separate atrioventricular rings, mitral valve cleft with regurgitation, and no significant VSD. Surgical repair was considered to be ideal in this case. In the operation of congenital heart disease, however, accurate anatomical information should be provided to improve the operative result. New anatomical findings would force the operator to change the surgical technique.

In the present case, three-dimensional echocar-

diography visualized morphology of the mitral valve cleft and ASD, being useful for classifying the type of AVSD. RT-3DE provided additional information for 12 of 33(36%)patients with congenital heart disease and enhanced diagnostic confidence.[6] Color Doppler three-dimensional echocardiography facilitated recognition of the existence and direction of shunt. In conclusion, transthoracic RT-3DE provided more precise anatomical insight in the present case of partial AVSD, but optimal image quality is needed.

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要　約

三次元心エコー図法によって観察された部分房室中隔欠損の1例

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幼少時より心雑音を指摘されていた48歳の女性が，息切れの増悪のため入院した．心エコー図法により心房中隔の一次口欠損と心室中隔上端の嚢状の構造物が認められ，カラードップラー法で心房中隔を主として拡張期に通過するシャント血流および僧帽弁前尖の亀裂からの僧帽弁逆流が指摘された．経胸壁からの三次元心エコー図法では僧帽弁前尖に三尖弁方向に向かう亀裂と，そこからの僧帽弁逆流，房室弁に隣接する短径14mmの心房中隔欠損が認められ，そこを通り心房壁後方に向かうシャント血流が認められ，部分型房室中隔欠損症と診断された．また，心室中隔の嚢状の構造物に欠損孔は伴ってなく，左室流出路狭窄や部分肺静脈灌流異常も認められなかった．三次元心エコー図法では解剖学的情報を立体的に把握可能であり，複雑先天性心疾患の診断には三次元心エコー図法が有用であることが示された．

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References

1) Bergin ML, Warnes CA, Tajik AJ, Danielson GK: Partial atrioventricular canal defect: Long-term follow-up after initial repair in patients ≧ 40 years old. J Am Coll Cardiol 1995; **25**: 1189−1194
2) Burke RP, Horvath K, Landzberg M, Hyde P, Collins JJ, Cohn LH: Long-term follow-up after surgical repair of ostium primum atrial septal defects in adults. J Am Coll Cardiol 1996; **27**: 696−699
3) Moran AM, Daebritz S, Keane JF, Mayer JE: Surgical management of mitral regurgitation after repair of endocardial cushion defects: Early and midterm results. Circulation 2000; **102**(Suppl Ⅲ): Ⅲ160−Ⅲ165
4) Miyamoto K, Nakatani S, Kanzaki H, Tagusari O, Kobayashi J: Detection of discrete subaortic stenosis by 3-dimensional transesophageal echocardiography. Echocardiography 2005; **22**: 783−784
5) Abe H, Nakatani S, Kanzaki H, Hasegawa T, Miyatake K: The structual and dynamic recognition of discrete subaortic stenosis by real-time three-dimensional transthracic echocardiography. J Echocardiogr 2005; **3**: 48−49
6) Salustri A, Spitaels S, McGhie J, Vletter W, Roelandt JR: Transthoracic three-dimensional echocardiography in adult patients with congenital heart disease. J Am Coll Cardiol 1995; **26**: 759−767