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Adult Left Isomerism Complicated With Life Threatening Arrhythmias Detected by the Use of Biplane Transesophageal Echocardiography : A Case Report

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Abstract

Left isomerism with critical arrhythmia in a 34-year-old housewife is reported. Left isomerism is rare among adult congenital disorders. The prognosis in childhood depends on intractable heart failure, but is unclear in adulthood. We describe an adult patient with left isomerism who recovered from life-threatening arrhythmias. Transesophageal echocardiography was especially useful to identify the morphology of both atrial appendages.

Key Words

congenital heart disease (left isomerism), ventricular tachycardia, cardiac pacing (artificial), echocardiography (biplane transesophageal)

INTRODUCTION

Left isomerism is a congenital disorder of the situs including three parts of thoracic, atrial and abdominal isomerism, and is frequently associated with various types of visceral anomalies, especially cardiac anomalies[1,2]. The prognosis of this disease in childhood depends on intractable heart failure due to associated cardiac anomalies, but remains unclear in adulthood[1,2]. Recently, sinus dysfunction[3] and atrioventricular block[4] have been well documented in this disorder. We experienced an adult patient with left isomerism who recovered from life-threatening arrhythmias. Transesophageal echocardiography was useful for delineating the morphology of the atrial appendages.

CASE REPORT

A 34-year-old housewife who had had two pregnancies and two deliveries was admitted to our hospital with gastric ulcer. Her history included spontaneous closure of a ventricular septal defect and significant bradycardia (40 /min) accompanied by occasional loss of consciousness. On the ninth hospital day, she suddenly developed syncope and convulsion. Electrocardiography showed ventricular fibrillation and tachycardia (**Fig. 1–A**). Following resuscitation, bradycardia of less than 40 /min persisted (**Fig. 1–B**) along with ectopic atrial rhythm (**Fig. 1–C**). Temporary right ventricular pacing was started but atrioventricular (A-V) junctional rhythm and A-V dissociation with significant bradycardia were also observed (**Fig. 1–D,E**). One week later, a permanent pacemaker was implanted and the pa-

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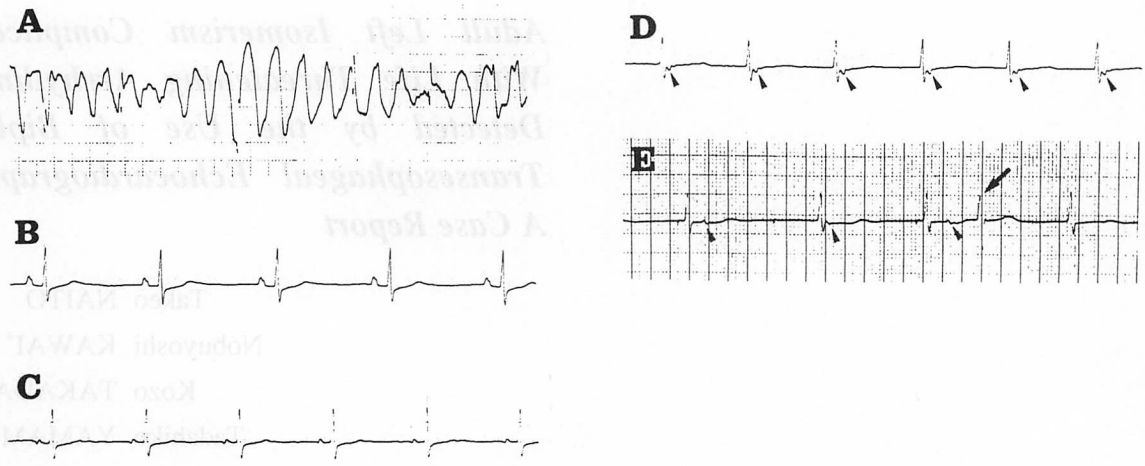

Fig. 1 Serial electrocardiograms

A : Torsades de pointes during syncopal attack after resuscitation.
B : Original sinus rhythm.
C : Biphasic P waves with different morphology to *B* and shortened PR interval.
D : Retrograde P wave (70/min) (*arrowheads*).
E : A-V dissociation, pacing artifacts without ventricular capture, slow atrial beats (*arrowheads*) and escaped beats and captured beat (*arrow*).
(electrocardiograms leads; panel *A* : monitoring lead, *B to E* : aVF)

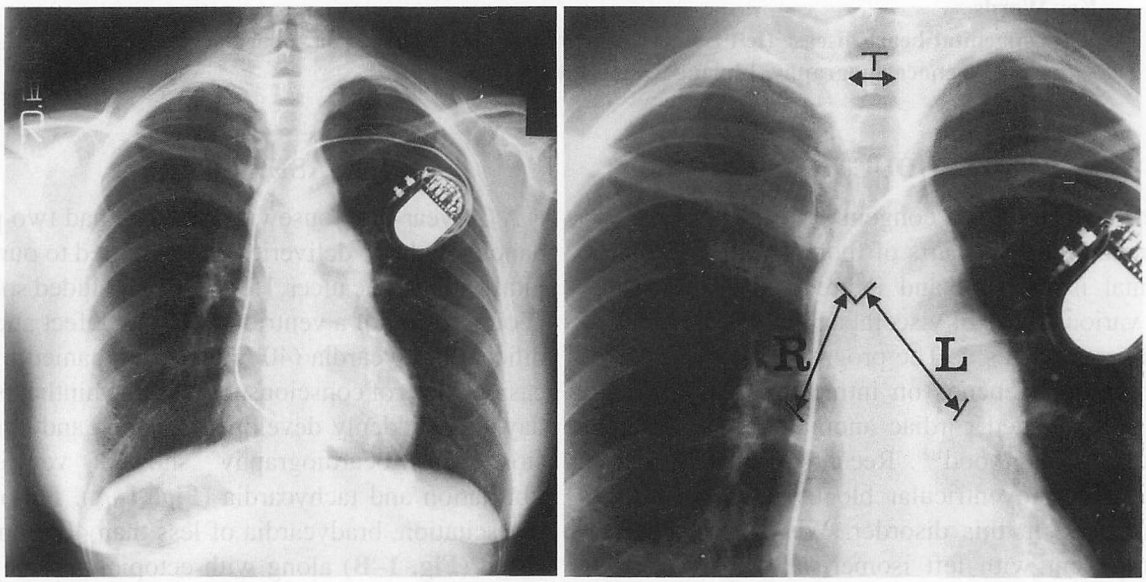

Fig. 2 Chest radiographs

Left : Postero-anterior view of the chest revealing marked enlargement of left second arch but no increase in the cardiothoracic ratio. The gastric bulb is located on the right.
Right : Magnified view of the bifurcation of the main bronchi. The bronchial length ratio (L/R) is less than 1.5, and left bronchial length (L/T) divided by tracheal width is more than 2.5[6,7].

tient became asymptomatic with a normally functioning pacemaker.

Chest radiography, measurement of the bronchial length ratio and division of the left bronchial length by tracheal width suggested thoracic left isomerism (**Fig. 2**). A transthoracic echocardiogram showed normal continuity from the traveculated left-sided ventricle to the aorta, three leaflets of the left-sided A-V valve and enlargement of the pulmonary trunk. The transesophageal echocardiogram showed bilat-

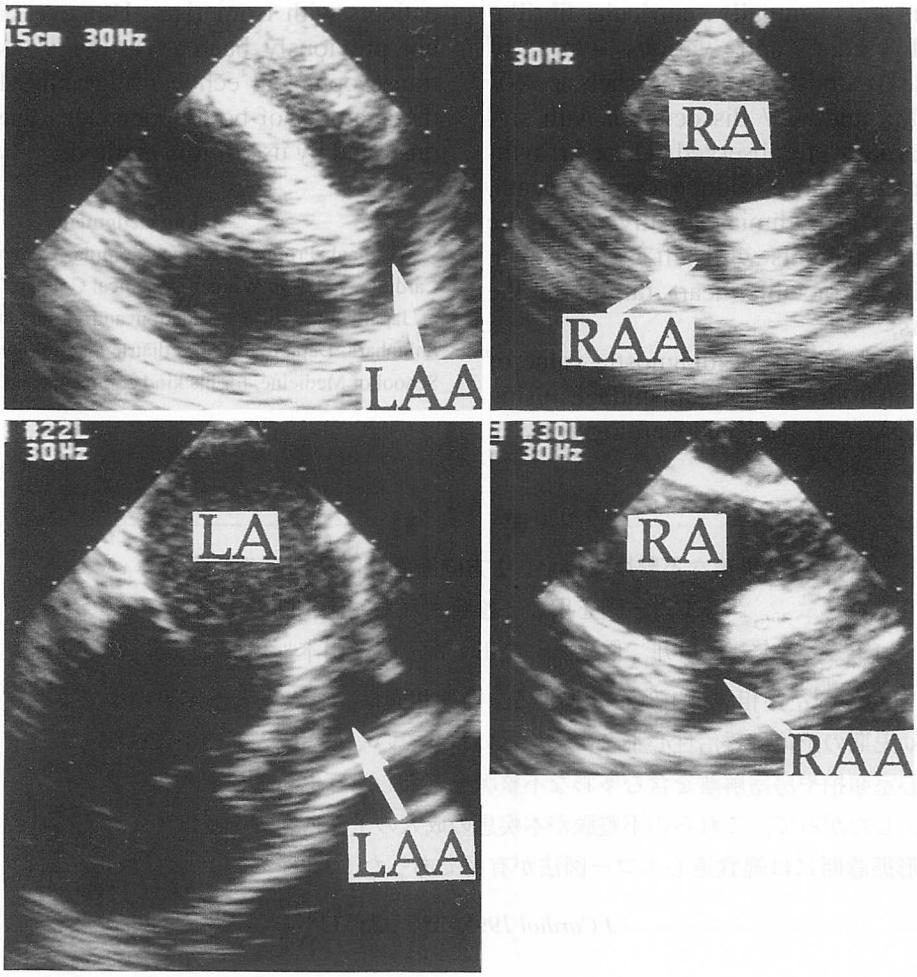

Fig. 3 Biplane transesophageal echocardiograms, transverse (*upper*) and longitudinal (*lower*) views
The left-sided atrial appendage (LAA) has a narrow connection to the atrium (LA) and a long cavity (*upper and lower left*). The right-sided atrial appendage (RAA) has a narrow connection to the atrium (RA) and a long cavity (*upper and lower right*).

eral morphological left atrial appendages (**Fig. 3**). These findings suggested atrial left isomerism. Cardiac catheterization revealed normal hemodynamic findings. A left-sided ventriculogram revealed morphological right ventricle. A coronary angiogram showed inverted normal coronary distribution and no stenotic lesion. A right-sided angiogram revealed interruption of the inferior vena cava, azygos continuation, and direct connection between the right atrium and hepatic vein. A right-sided ventriculogram revealed morphological left ventricle, slightly depressed mobility of the pulmonic valve, and enlarged pulmonary trunk. These findings suggested corrected transposition of the great arteries. Abdominal computed tomography revealed polysplenia, malrotation of the intestine, and right-sided stomach.

DISCUSSION

Left isomerism in adult is a rare congenital disorder. Peoples *et al*[1] reported three adult cases of left isomerism associated with the corrected transposition of the great arteries in 142 autopsy cases. The prognosis of left isomerism in adults is still unknown. Ten percent of patients survive until mid-adolescence[1,2]. The most common cause of death is intractable heart failure due to associated cardiac anomalies[1,2], but the importance of arrhythmias in prognosis is not fully understood. Recently, the characteristics of sinus function in cardiac left isomerism have been investigated showing the number of patients with slow atrial rate increased with age[3]. In addition, A-V block is also associated with this disorder[4]. In our case, bradycardia had been present

for several years and eventually ventricular fibrillation and tachycardia, and various types of arrhythmias due to sinus dysfunction such as A-V junctional rhythm and A-V dissociation with significant bradycardia were observed. These arrhythmias could directly cause sudden cardiac death. Therefore, serious arrhythmias greatly affect the prognosis in adult patients with left isomerism in comparison with intractable heart failure in the younger patients.

Stümper *et al*[5] reported the diagnostic value of delineating the anatomy of both appendages using single-plane transesophageal echocardiography in patients with isomerism. However, isomerism has not previously been investigated using "biplane" transesophageal echocardiography. In our patient, the anatomy of both appendages was clearly demonstrated by use of this method.

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要　約

重篤な不整脈を伴い，2方向断面探触子による経食道心エコー図を用いて診断された左側相同の1例

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34歳，女性の左側相同の1例を報告する．左側相同は，成人先天性疾患の中でまれな病態である．幼児期の予後は難治性心不全によるといわれているが，成人での予後は明らかでない．本症例は心室頻拍や房室解離を含む多彩な不整脈をきたし，蘇生および永久ペースメーカーを必要とした．したがって，これらの不整脈が本疾患の成人の予後に影響を及ぼすと考えられた．また心耳の形態診断には経食道心エコー図法が有用であった．

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