

Long-term Survival of Patients With Unoperated Single Ventricle Heart Defect: Four Case Reports and Literature Review

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Case report

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Abstract

Background: A single ventricle heart defect is a rare complex congenital cardiac malformation. It is rare for patients with single ventricle to spontaneously survive long-term to adulthood. No literature was retrieved that summarized the ultrasonographic features of patients with single ventricle who survived for a long time without surgery.

Case presentation: Four patients in this study were diagnosed with single ventricle heart defects during childhood, but received no surgical intervention. All four patients had good outcomes. We summarize the ultrasound findings of these patients.

Conclusion: We summarize the factors that contribute to the long-term survival of single ventricle patients without surgical intervention, and discuss the diagnosis and treatment of single ventricle to improve the survival rate of patients.

Introduction

A single ventricle (SV) heart defect is a rare complex congenital cardiac malformation, accounting for approximately 1%-2% of congenital heart diseases.¹ The SV can be distinguished as having a left ventricle (LV) or right ventricle (RV) morphology, and in rare cases, the morphology is indeterminate.² Patients with SV have only one functional ventricular chamber, and blood from the body and pulmonary circulation is mixed in the heart chamber, often causing overload in the single functional ventricular chamber and leading to heart failure in the early stages.³ Surgical intervention is the main treatment for SV patients. In this study, we report four adult patients who had SV since birth without surgical intervention and survived to adulthood. All the case information and the images (Figs. 1–4) in this review have been approved by the ethics committee of our hospital. We also conducted a Medline search and reviewed similar cases published in recent years.

Case Presentations

Case 1

A 13-year-old female patient (height: 150 cm; weight: 42 kg) was diagnosed with functional SV combined with transposition of the great arteries (TGA) with a right position of the aorta based on ultrasound findings at the age of 3, but did not receive any treatment. In 2017, she came to our hospital due to numbness in both lower limbs and dyskinesia. Thoracic magnetic resonance imaging (MRI) showed a space-occupying lesion in the spinal canal. A bone biopsy was performed and revealed cavernous hemangioma. This patient underwent a tumor resection at another hospital and returned to our hospital for radiotherapy 5 months after the operation. Physical examination at our hospital showed a temperature of 36.5°C, a pulse of 80 beats/min, no difference in the pulse and heart rate (HR), a respiratory rate of 18 breaths/min, and a blood pressure of 120/80 mmHg. Other pertinent examinations showed no signs of

anemia and she had no history of hemoptysis, syncope, or long-term high fever. The patient exhibited normal development, moderate nutrition. Her New York Heart Association functional class was II. Echocardiography revealed the following: the heart was located in the left thoracic cavity, the atrium was in the normal position, the left ventricular cavity was the main cavity, and the right ventricular cavity was the residual cavity. The pulmonary artery (PA) located on the posterior side and arose from the left cavity. The aorta was moderately developed, located on the anterior side and arose from the right cavity. On Echocardiography, the upper interventricular septum was interrupted by 22.3 mm (Fig. 1). Color Doppler flow imaging (CDFI) revealed a two-way low-resistance shunt in the ventricular septal defect. The patient was diagnosed with the following diseases based on the echocardiography results: congenital heart disease, functional SV, TGA, mild PS, left ventricular hypertrophy, and left atrial enlargement. Further examination was recommended. No tumor metastasis was found after radiotherapy at our hospital and the prognosis was appreciable.

Case 2

A 41-year-old female patient (height: 163 cm, weight: 50 kg) was admitted to our hospital due to chest pain, chest tightness, and palpitations lasting for 3 days. She presented with cyanosis of the mouth and limb immediately after birth, with obvious cyanosis after crying but without syncope or squatting signs. She was diagnosed with a congenital heart disease at the local hospital but was not treated. Her growth and development were not affected and daily activities were only slightly limited. After catching a cold 3 days earlier, she experienced chest tightness and shortness of breath after general physical activities. After symptomatic treatment at the local hospital, she was admitted to our hospital due to a lack of obvious symptom relief.

Physical examination at our hospital revealed a temperature of 36.6°C, a pulse of 62 beats/min, a respiratory rate of 20 breaths/min, a blood pressure of 110/60 mmHg, clear consciousness, no yellowing of the skin and sclera, and cyanosis of the mouth. Her heart rate was 62 beats/min, the heart rhythm was uniform. Her New York Heart Association functional class was I. Echocardiography showed dextrocardia as well as a heart with a functional single atrium and SV, ventricular hypertrophy, and a common atrioventricular valve. The atrial and ventricular septa were not explored but were clearly defined. Two large arteries, the PA and the aorta, were concomitant, both starting from the functional SV. The PA was located on the anterior side and the aorta was located on the posterior side. No obvious abnormalities were found in the aortic valve. CDFI revealed severe reflux in the common atrioventricular valve in the systolic stage, the neck of the valve orifice reflux was 13.4 mm, the flow rate was 4.0 m/s, and the pressure gradient (PG) was 63 mmHg (Fig. 2). Under three-dimensional ultrasonic examination, the atrioventricular valves appeared to consist of four valves. The echocardiographic diagnosis for this patient was congenital heart disease complex deformity, dextrocardia, single atrium, SV, PS, and severe common atrioventricular valve insufficiency. The patient was discharged after 5 days of anti-infection and symptomatic treatments. After discharge, we continued to follow up with the patient by telephone and outpatient visits.

Case 3

A 13-year-old female patient (height: 137 cm; weight: 40 kg) came to our hospital due to palpitations and chest tightness lasting for 2 days. She was diagnosed with a complex congenital heart disease at the age of 6 months but was not treated. Physical examination on admission showed a body temperature of 36.4°C, a pulse of 57 beats/min, a respiratory rate of 21 breaths/min, and a blood pressure of 105/70 mmHg. Her New York Heart Association functional class was II. Echocardiography showed the following: the heart was located in the left thoracic cavity, the atrium was in the normal position, the PA and the aorta were concomitant, and the PA was located in front of the aorta, both starting from a functional SV. The right atrium was enlarged, no atrioventricular valve was found in the left ventricular cavity parallel to the right atrium, the echo of the interventricular atrium was interrupted by 10.2 mm, and the internal diameter of the SV was 65 mm. CDFI showed a shunt in the atrial septal defect with a flow rate of 2.6 m/s and a PG of 29 mmHg. There were severe reflux in the atrioventricular valve in the systolic stage, the regurgitation area of the atrioventricular valve orifice was 3.7 cm², the flow rate was 5.4 cm/s, and the PG was 117 mmHg (Fig. 3). The length of the pulmonary valve regurgitation beam was 22.8 mm, the flow rate was 3.5 m/s, the PG was 50 mmHg, and the average PA pressure was estimated to be 55 mmHg. The echocardiographic diagnosis was congenital heart disease (SV, atrial septal defect), mild atrioventricular valve insufficiency, mild pulmonary valve regurgitation, and pulmonary hypertension (PHT). The patient was discharged after 3 days of symptomatic treatment.

Case 4

A 39-year-old woman (height: 162 cm; weight: 50 kg) came to our hospital for chest pain and chest tightness lasting for 4 days. She was diagnosed with a congenital heart disease at birth but was not treated. Physical examination on admission showed a temperature of 37.0°C, a pulse of 60 beats/min, a respiratory rate of 22 breaths/min, and a blood pressure of 110/80 mmHg. Her New York Heart Association functional class was II. Echocardiography showed the following: the heart was located in the left thoracic cavity, the atrium was in the normal position, the positions of the PA and the aorta were abnormal, the aorta had widened and moved forward, and both the heart and PA started from the functional SV. There was no ventricular septum, the internal diameter of the SV was 58.2 mm (Fig. 4). CDFI revealed that the regurgitation area of the tricuspid valve orifice was 3.1 cm², the flow rate was 4.1 m/s, and the PG was 66 mmHg. The echocardiographic diagnosis for this patient was congenital heart disease (SV, left ventricular type, PHT, and mild tricuspid regurgitation). The patient was discharged after 4 days of symptomatic treatment.

Discussion

SV heart defect is a rare congenital cyanotic heart disease often associated with PS, TGA, atrial septal defect, and other malformations. Echocardiography is the first choice for the diagnosis and evaluation of SV. Echocardiography can also be used to diagnose cardiac malformations in the early and second trimester of pregnancy, to make accurate judgments regarding the classification of SV, its associated malformations, and atrioventricular valve openings and reflux, as well as to evaluate ventricular function.⁴ SV heart defects can be divided into four types according to the morphological structure: Type

A, the left ventricular main chamber is dominant; Type B, the right ventricular main chamber is dominant; Type C, the ventricular septum is absent, and the left and right ventricular structures constitute the ventricular cavity; and Type D, indeterminate.⁶ Unlike echocardiography, three-dimensional ultrasound can visually display the internal structure of the heart, thus improving the accuracy of preoperative diagnosis, and is of great significance in the selection of surgical methods and the assessment of prognosis.⁷ We reviewed literature reports of SV heart defect cases involving patients over 13 years of age without surgical intervention and summarized the common characteristics that were correlated to the long-term survival of the patients (Table 1). The aforementioned four patients in this study were diagnosed with SV heart defects during childhood, but received no surgical intervention. All four patients had good outcomes. Table 1 shows that the clinical manifestations and long-term prognosis of patients with SV heart defects primarily depend on the presence or absence of pulmonary blood flow obstruction, pulmonary vascular resistance, and ventricular shape and function. Whether the aortic blood flow is blocked and whether the shape and function of atrioventricular valve still function play an important role in patient prognosis. Moderate PS is critical for hemodynamic balance in SV patients; first, it restricts pulmonary blood flow and prevents pulmonary arteriole disease; second, it restricts the left-to-right shunt, prevents pulmonary blood flow increase and ventricular overload, and maintains sufficient pulmonary blood flow to supply oxygen. In addition, SV patients with adequate oxygenation and balanced ventricular load may have a chance for long-term survival. The morphology and function of the ventricles is another important factor. There are, in fact, no reports of long-term survival for patients with right morphology or an indeterminate-type SV. This is probably due to the intrinsic capacity of the left ventricle to deal with systemic resistance. When all other conditions are equal, patients with a single left ventricle heart defect have better prognosis than those with a single right ventricle heart defect. When aortic blood flow resistance increases, the ventricular afterload also increases, leading to a low cardiac output, which is not conducive to survival. In addition, effective and detached atrioventricular valves have a positive effect on survival. Atrioventricular valve dysfunction leads to ventricular volume overload. The optimal anatomical structure for SV patients is a normal morphology for the left ventricle, great artery displacement, no systematic obstruction of outflow tract, full atrioventricular valve function, and moderate PS. These anatomical features enable patients to survive mild to moderate symptoms and enjoy a good quality of life. It should also be noted that arrhythmias are the most common complications in older SV patients.⁸ With the improvement of surgical techniques and postoperative care, more SV patients have been able to survive for a longer period of time. The main treatment methods for SV patients are radical surgery, palliative surgery, and heart transplantation. The palliative surgery mainly includes modified Blalock-Taussig (B-T) shunt operation, PA banding, Norwood operation, Glenn operation, and Fontan operation.⁹

Table 1
Reported unoperated SV patients with an age over 13 years

References	Published time	Age /Sex	Anatomy	DILV	NYHA class	PHT	Arrhythmias
Warner et al ¹⁰	1996	64/F	TGA	Yes	-	Yes	1°AVB
Gabbarini et al ¹¹	1999	61/M	MGA	Yes	II-III	Yes	Paroxysmal atrial fibrillation
Hager et al ⁴	2002	62/F	TGA PS	Yes	III	Yes	Atrial fibrillation
Restaino et al ¹²	2004	57/F	TGA	Yes	II	Yes	-
Book et al ¹³	2007	71/F	TGA	Yes	-	Yes	NSR
Park et al ¹⁴	2007	41/M	TGA	Yes	II-III	Yes	Atrial fibrillation
Coats et al ¹⁵	2010	59/F	MGA	Yes	III	Yes	Atrial flutter
Paul et al ¹⁶	2012	36/M	MGA	Yes	IV	Yes	Atrial fibrillation
Wang et al ¹⁷	2013	35/F	MGA	Yes	II	Yes	NSR
Gesuete et al ¹⁸	2016	74/M	TGA	Yes	II	Yes	Atrial flutter
Güvenç et al ¹⁹	2016	31/M	TGA	Yes	III	Yes	NSR
Alpat et al ²⁰	2017	45/M	TGA	Yes	-	Yes	Atrial fibrillation
Kongunattan et al ²¹	2017	27/M	TGA	Yes	-	No	-
Andrade et al ²²	2019	62/M	TGA	Yes	-	Yes	-
Present case 1	2020	13/F	TGA	Yes	II	Yes	NSR
Present case 2	2020	41/F	PS Single atrium	Yes	III	Yes	NSR
Present case 3	2020	13/F	Atrial septal defect	Yes	II	Yes	NSR

M = male; F = female; DILV = double inlet left ventricle; PHT = pulmonary hypertension; PS = pulmonary stenosis; MGA = ventriculoarterial concordance with left/right position of the aorta; TGA = transposition of the great arteries with left/right position of the aorta; AVB = atrioventricular block; NSR = normal sinus rhythm

References	Published time	Age /Sex	Anatomy	DILV	NYHA class	PHT	Arrhythmias
Present case 4	2020	39/F	TGA	Yes	III	Yes	NSR
M = male; F = female; DILV = double inlet left ventricle; PHT = pulmonary hypertension; PS = pulmonary stenosis; MGA = ventriculoarterial concordance with left/right position of the aorta; TGA = transposition of the great arteries with left/right position of the aorta; AVB = atrioventricular block; NSR = normal sinus rhythm							

Conclusion

With the development of new surgical methods for the treatment of SV heart defects, it is important to understand the long-term outcomes for patients who have not been operated on. While the Fontan operation has become a common and effective method for SV treatment, long-term outcomes are fraught with morbidity and mortality. The majority of patients in our series and in the literature to demonstrate advanced survival had DILV and PS. In our opinion, such patients with balanced hemodynamic condition should be followed and treated conservatively. Major cardiac surgery which leads to gross hemodynamic adjustments should be avoided. The optimal timing for surgery and appropriate surgical method can be determined based on: 1) the patient's symptoms and specific diagnosis of SV heart defect, and 2) the following conditions: the anatomy of the parting and concurrent heart malformations, the development of the PA, the amount of pressure and resistance, and ventricular function, with a significant aim to reduce mortality and improve the prognosis as well as quality of life of SV patients.

Declarations

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Disclosures

The Author(s) declare(s) that there is no conflict of interest.

Ethical statements

This article does not contain any studies with human or animal subjects performed by the any of the authors.

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Figures

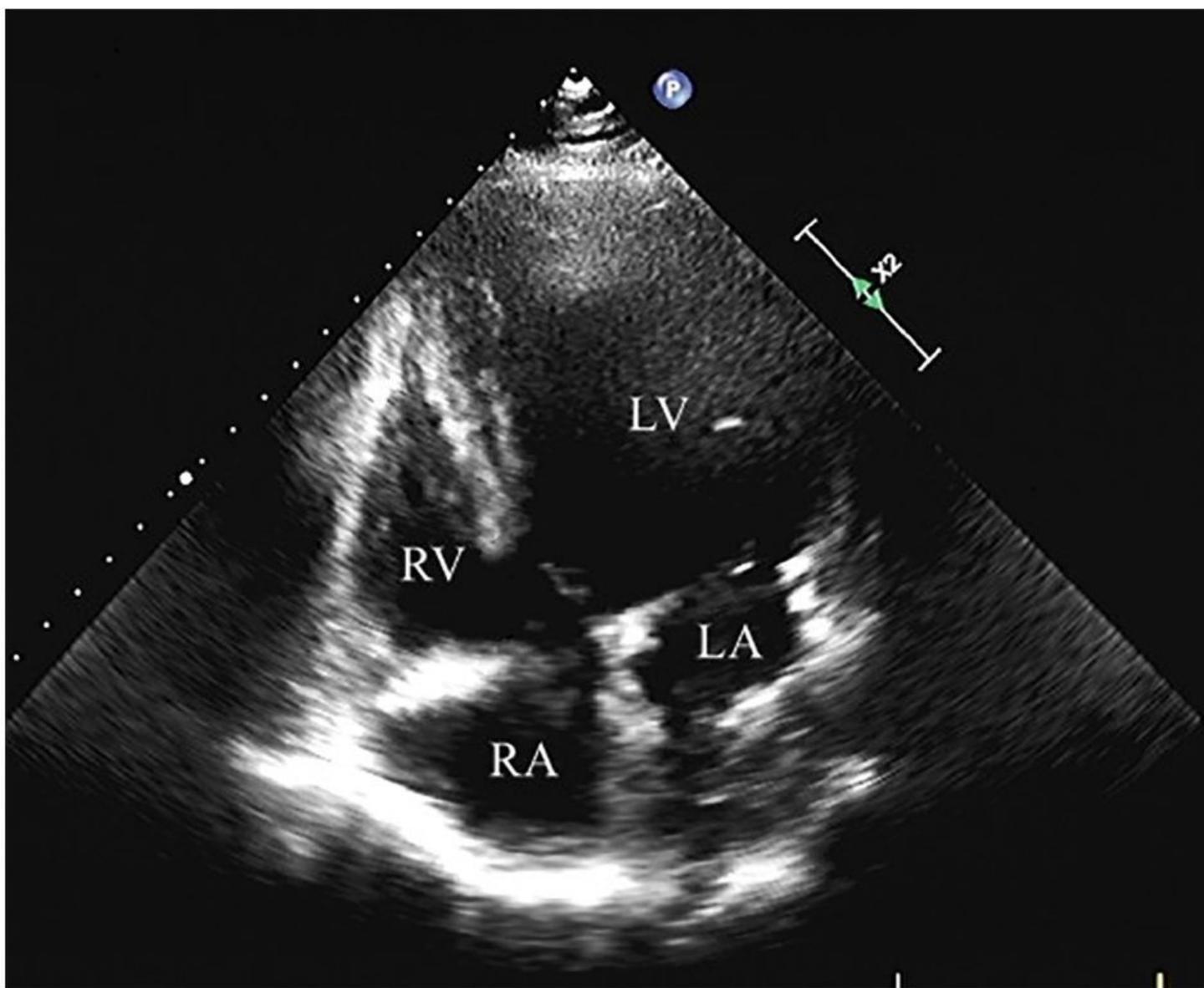


Figure 1

A 13-year-old female SV patient combined with cavernous hemangioma. Two-dimensional echocardiography: four-cavity cardiac surface view showing the left ventricular cavity was the main cavity, and the right ventricular cavity was the residual cavity. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

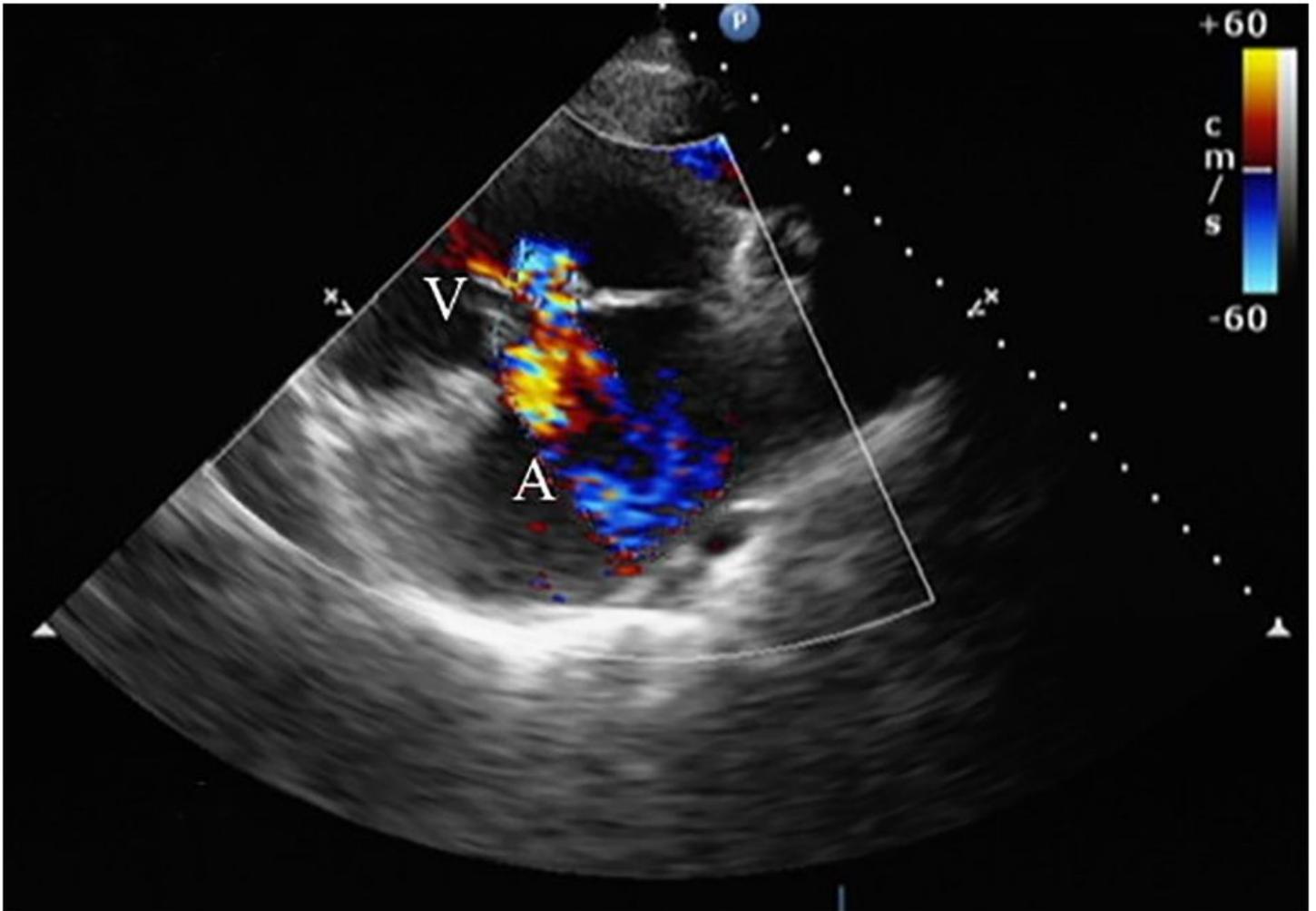


Figure 2

A 41-year-old female SV patient combined with single atrium. Two-dimensional echocardiography: four-cavity cardiac surface view showing a functional single atrium and a SV; CDFI showing severe reflux in the common atrioventricular valve in the systolic stage. A, atrium; V, ventricle

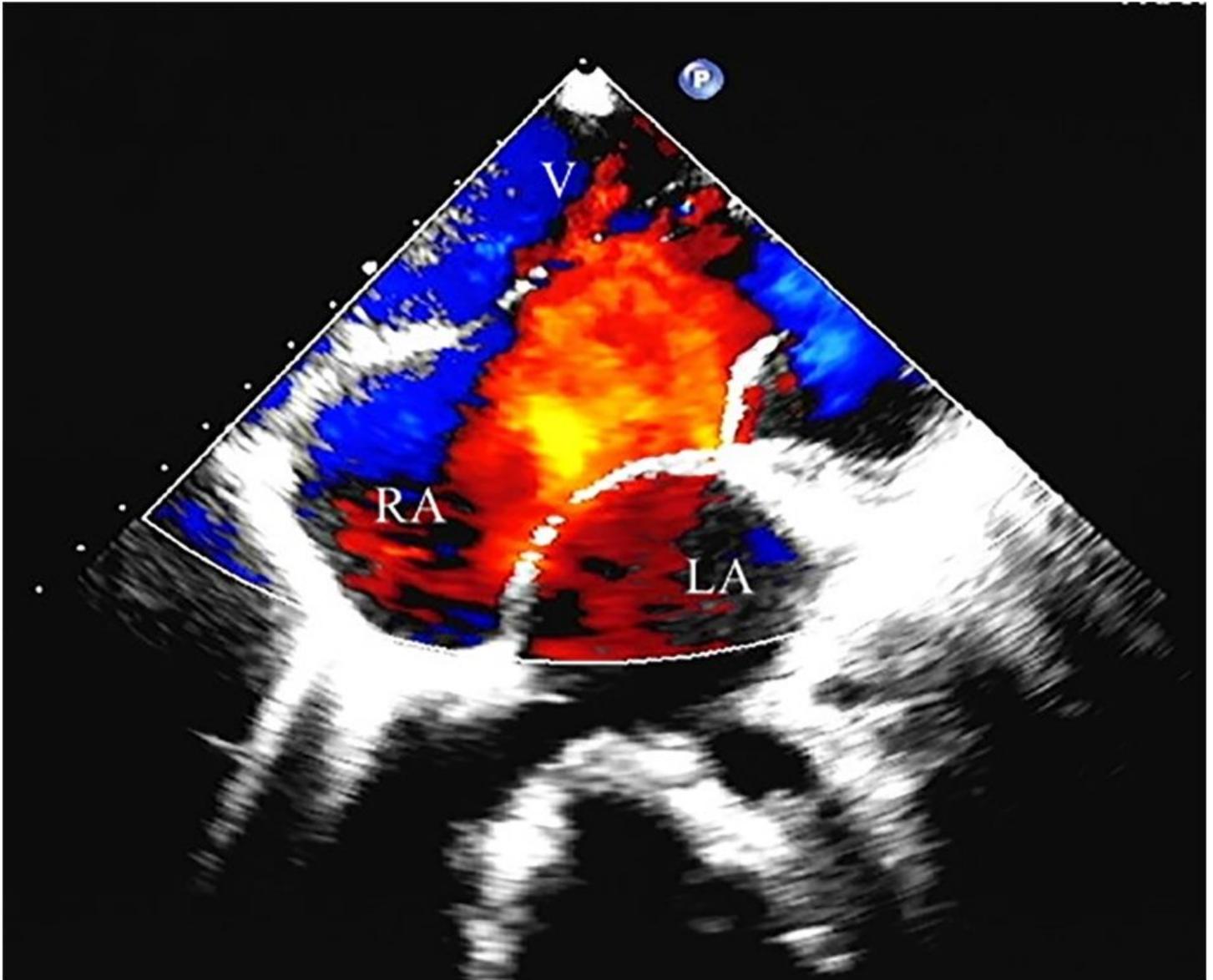


Figure 3

A 13-year-old female SV patient combined with atrial septal defect. A, Two-dimensional echocardiography: four-cavity cardiac surface view showing the right atrium was enlarged, no atrioventricular valve was found in the left ventricular cavity parallel to the right atrium. B, CDFI showing severe reflux in the atrioventricular valve in the systolic stage. LA, left atrium; RA, right atrium; V, ventricle

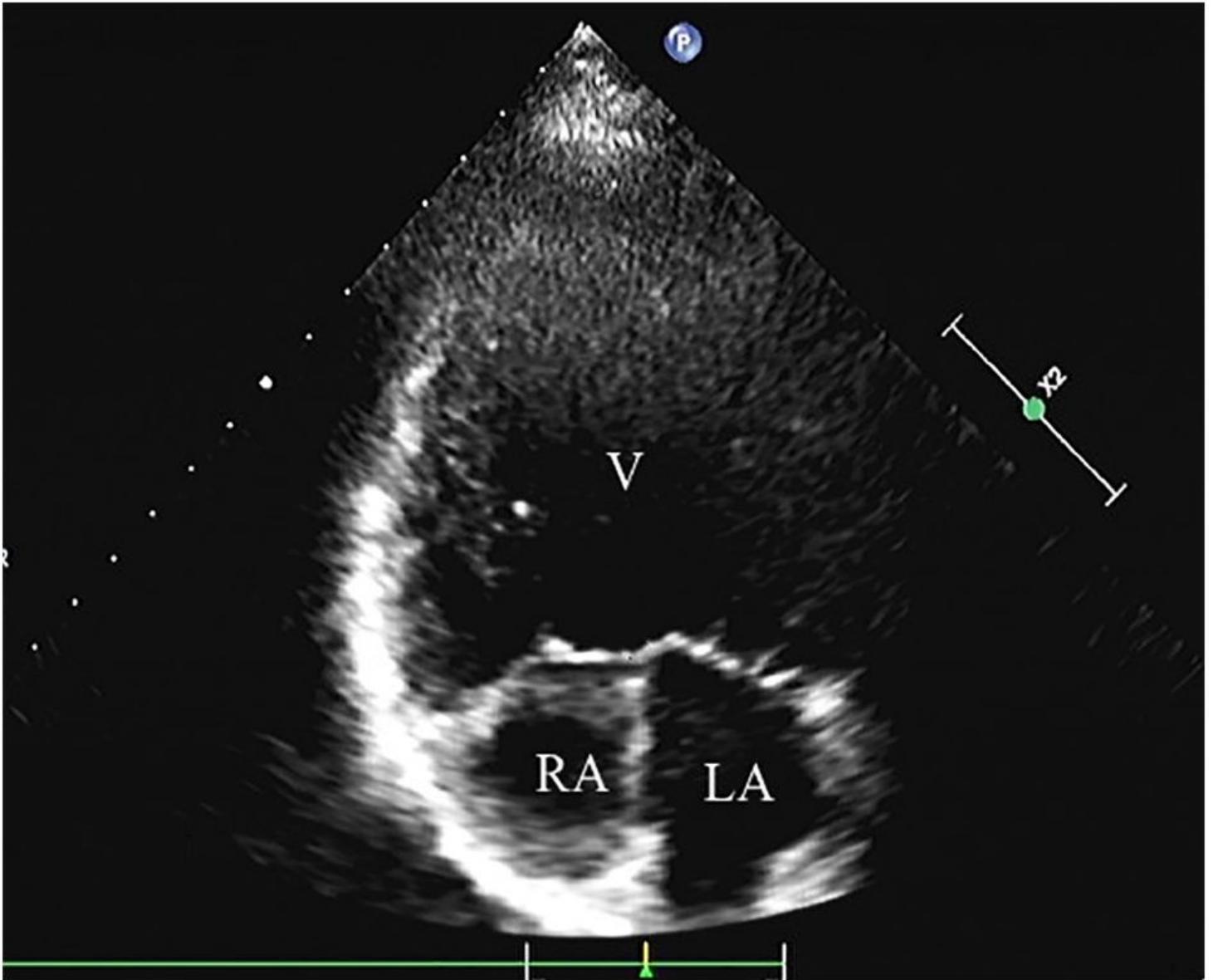


Figure 4

A 39-year-old female SV patient combined with patent ductus arteriosus. Two-dimensional echocardiography: four-cavity cardiac surface view showing two normal-sized atriums and an SV. LA, left atrium; LV, left ventricle; V, ventricle