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Single Ventricle Complicated by Atrial Arrhythmia in Adulthood: A Case Report and Review of the Literature

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Abstract **Case Report**

Background: Single ventricle (SV) is a rare congenital heart disease in which a single functional ventricular chamber supports both systemic and pulmonary circulations. Arrhythmias are frequent late complications and significantly impact prognosis. ***Case Presentation:*** We report a 29-year-old patient with SV physiology admitted for palpitations secondary to atrial flutter with variable conduction. Transthoracic echocardiography revealed a single right ventricle with double inlet, moderate-to-severe atrioventricular valve regurgitation, and pulmonary arterial hypertension. Clinical evolution was favorable under medical therapy, including beta-blockers, digoxin, anticoagulation, and diuretics. ***Conclusion:*** This case highlights the complexity of long-term follow-up in adults with SV physiology and emphasizes the importance of early recognition and management of arrhythmias to improve outcomes.

Keywords: single ventricle, atrial flutter, congenital heart disease, arrhythmia, adult.

INTRODUCTION

Single ventricle represents a heterogeneous group of congenital heart diseases characterized by a single functional ventricular chamber supporting both systemic and pulmonary circulations [1,2]. Approximately 70% of cases have a dominant left ventricle, while right ventricular morphology is more commonly observed in double inlet ventricles, tricuspid atresia, and hypoplastic left heart syndrome [2,3].

Advances in surgical palliation and medical care have improved survival, leading to an increasing population of adults living with SV physiology [3]. However, these patients remain at high risk for long-term complications, particularly arrhythmias, which are a major cause of morbidity and mortality [4,5].

We report a case of atrial flutter in an adult with SV physiology and review the literature on arrhythmia management, risk factors, and long-term outcomes [6,7].

CASE PRESENTATION

A 29-year-old patient with a history of single ventricle physiology since birth, who had undergone pulmonary artery banding during childhood, was admitted to our department for progressive-onset palpitations. The patient had previously been hospitalized in 2021 for cardiac decompensation secondary to pneumonia.

On physical examination, the patient was conscious and hemodynamically stable, with a heart rate of 155 beats per minute, blood pressure of 120/80 mmHg, and oxygen saturation of 92% on room air. Cardiovascular examination revealed a median sternotomy scar, a loud second heart sound at the pulmonary area, and a systolic murmur radiating to all auscultatory foci.

Electrocardiography showed right axis deviation with atrial flutter and variable atrioventricular conduction, with a mean ventricular rate of approximately 150 bpm. Chest radiography demonstrated cardiomegaly with a subdiaphragmatic apex and straightening of the left middle cardiac border. Laboratory investigations, including thyroid function tests, were within normal limits.

Transthoracic echocardiography revealed a univentricular heart of right ventricular morphology with double inlet and preserved systolic function. The atrioventricular valves were nearly aligned with moderate-to-severe regurgitation. There was vascular malposition with a very anterior aorta and posterior pulmonary artery. The pulmonary artery band was

deemed loose with associated pulmonary arterial hypertension. The left atrium was dilated, while pulmonary and systemic venous returns were normal.

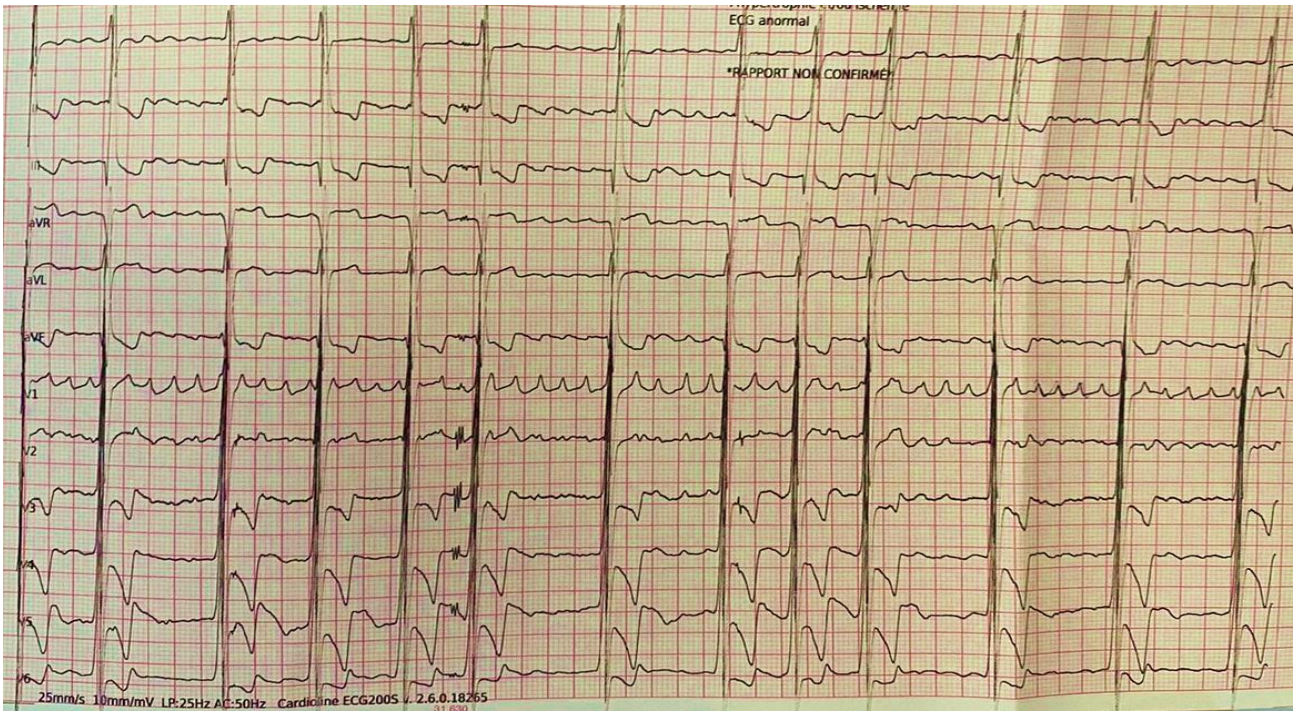

Figure 1: Electrocardiogram showing right axis deviation with atrial flutter and variable atrioventricular conduction, with a mean ventricular rate of approximately 150 beats per minute

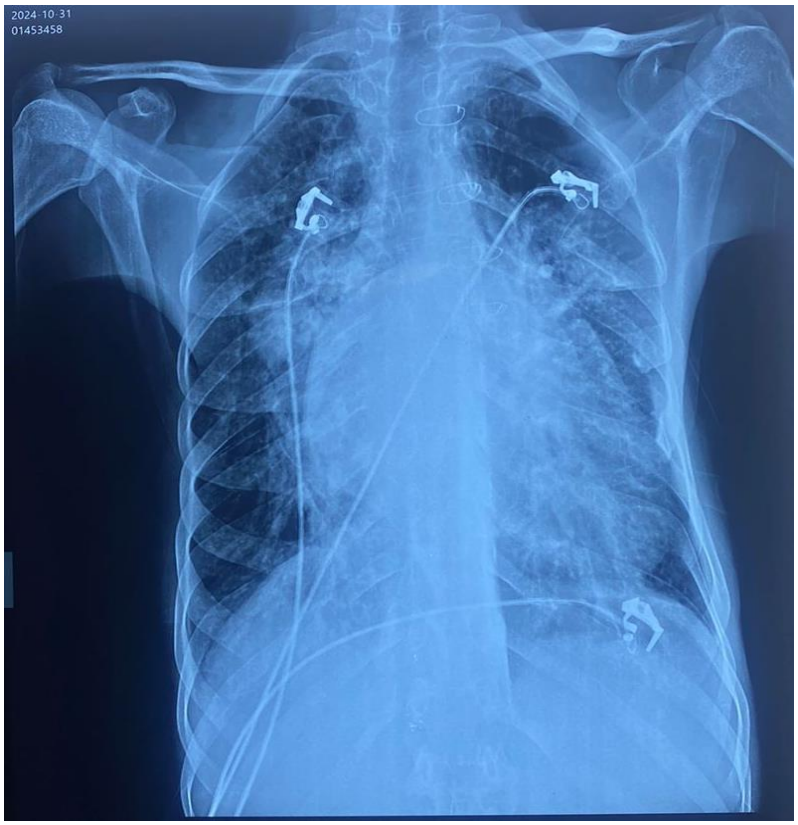

Figure 2. Chest X-ray demonstrating cardiomegaly with a subdiaphragmatic cardiac apex and straightening of the left middle cardiac border

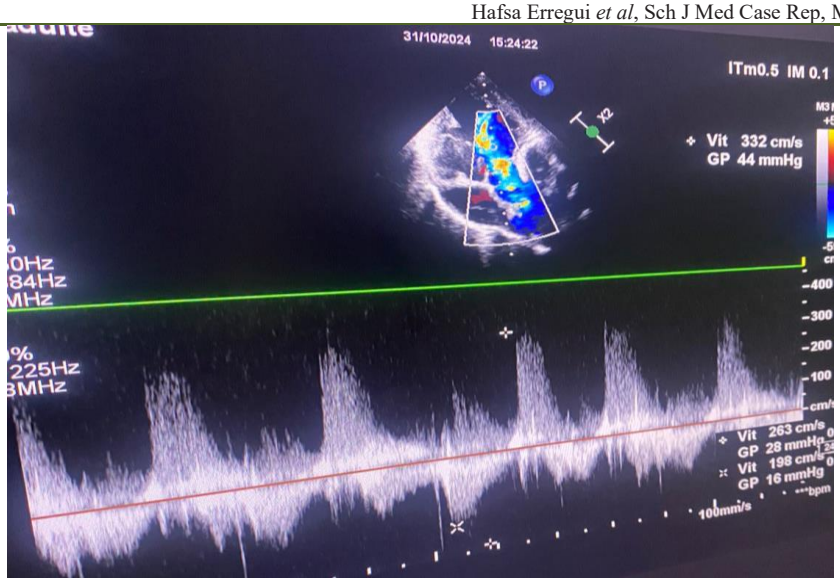

Figure 3: Transthoracic echocardiography showing pulmonary artery flow with a loose pulmonary artery band and features of pulmonary arterial hypertension

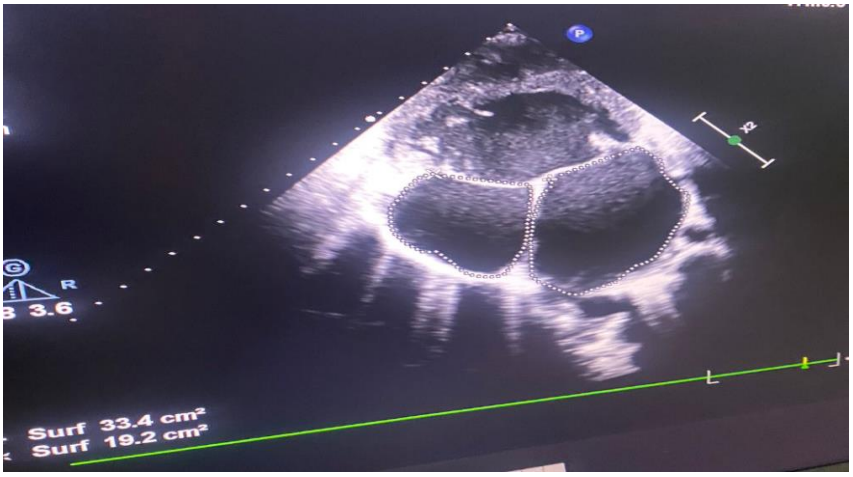

Figure 4. Apical four-chamber view demonstrating a single right ventricle with dilated atria

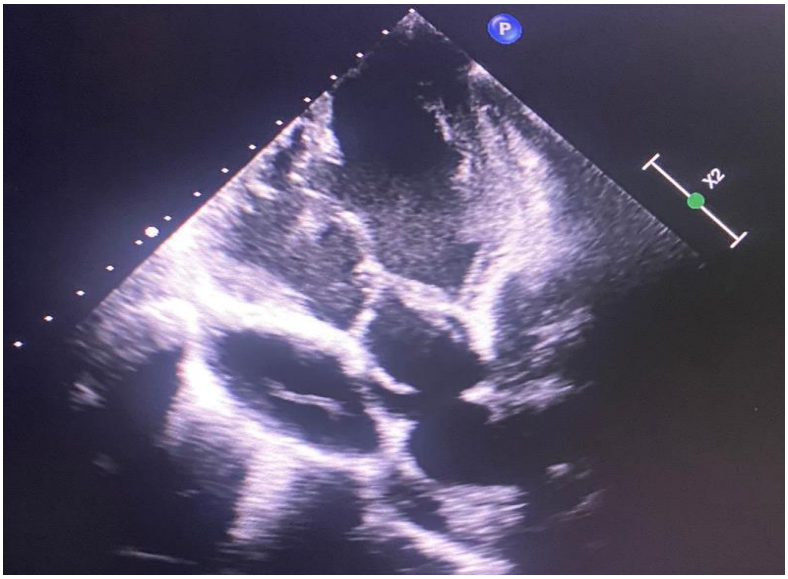

Figure 5. Apical echocardiographic view showing a single ventricle of right ventricular morphology with double inlet and nearly aligned atrioventricular valves

The patient was treated medically with beta-blockers, digoxin, anticoagulation, diuretics, and an aldosterone antagonist, with good clinical and hemodynamic evolution.

DISCUSSION

Adults with SV physiology represent one of the most complex populations within adult congenital heart disease. Arrhythmias, particularly atrial flutter and atrial fibrillation, are among the most frequent late complications and are associated with increased morbidity, heart failure hospitalizations, thromboembolic events, and mortality [4,5,11].

Mechanisms of arrhythmogenesis

include atrial dilation, surgical scars, abnormal myocardial architecture, chronic volume/pressure overload, and progressive ventricular dysfunction [6,7,12]. Arrhythmias may cause rapid hemodynamic deterioration due to limited preload reserve and dependence on atrial contribution to ventricular filling.

MANAGEMENT

is individualized, incorporating rhythm/rate control, long-term anticoagulation, and, in selected cases, catheter ablation or device therapy. Digoxin has been associated with improved survival in adults with SV physiology, although mechanisms remain unclear [8,9].

This case highlights the importance of early recognition and prompt management of arrhythmias in adults with SV physiology, as well as the need for structured lifelong follow-up in specialized congenital heart disease centers [3,4,11].

CONCLUSION

Single ventricle physiology is a complex congenital heart condition associated with significant long-term morbidity. Supraventricular arrhythmias represent a major prognostic factor and require early diagnosis and tailored management. Multidisciplinary care and long-term specialized follow-up are essential to improve survival and quality of life in this growing adult population.

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