ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.ijcdw.org

Indian Journal of Cardiovascular Disease in Women

Case report *Cardiovascular*

A Tale of Intricate Connections: An Interesting Case of Congenital Heart Disease with Supraventricular Tachycardia

Imamuddin Syed[1], Ravi Srinivas[1], M. U. Balachander[1], Praveen Nagula[1]

[1]Department of Cardiology, Osmania Medical College/Osmania General Hospital, Hyderabad, Telangana, India.

***Corresponding author:**
M. U. Balachander,
Department of Cardiology,
Osmania Medical College/
Osmania General Hospital,
Hyderabad, Telangana, India.

dr.balachander1404@gmail.com

Received: 25 June 2023
Accepted: 17 July 2023
Epub Ahead of Print : 04 August 2023
Published: 16 December 2023

DOI
10.25259/IJCDW_47_2023

Quick Response Code:

ABSTRACT

Double inlet left ventricle (DILV) is a rare complex congenital heart defect characterized by the presence of two atrioventricular valve orifices leading into a single left ventricle. Patients rarely survive beyond teenage years without surgical correction. Supraventricular tachycardia (SVT) in DILV is a rare occurrence that adds an additional layer of complexity to diagnosis and management. Here, we report an exceptional case of DILV who survived well into her fourth decade of life without any surgical correction. The patient presented to us with SVT which lead to the discovery of her unique cardiac anomaly.

Keywords: Complex congenital heart disease, Double inlet left ventricle, Supraventricular tachycardia

INTRODUCTION

Double inlet left ventricle (DILV) is a form of functionally univentricular heart defined by atrioventricular (AV) connection, in which the morphologically left ventricle (LV) receives more than 50% of the AV valves when they are separated or more than 75% of the common AV valve.

DILV is a rare congenital heart defect, with a prevalence of 5–10/100,000 population[1] and accounting for 1–5% of all congenital heart diseases. Precise nationwide statistics specific to DILV in India are not readily available.

In the largest series of patients with a single ventricle without surgical treatment, it was found that in the spectrum of DILV most patients die in their late teens to early 20s.[2-4] The common causes of death in patients without surgery include congestive heart failure, arrhythmias, or sudden death.

CASE REPORT

A 34 year female, born out of non-consanguineous marriage, had a history of New York Heart Association Class II dyspnea since the age of 14 years for which she was not evaluated. She was diagnosed to have heart disease at the age of 21 years during prenatal evaluation of her first pregnancy. She was advised to continue the pregnancy and had a normal vaginal delivery with no complications.

The patient presented to us with palpitations over the past 24 h. On examination, there was no cyanosis or clubbing. She had engorged neck veins (Frog sign). Her heart rate was 210 bpm and her blood pressure was 100/70 mm Hg. SpO_2 on room air was 95%. She had a pansystolic murmur at the left parasternal area and an ejection systolic murmur at the pulmonary area. Other systems examination was normal.

The electrocardiogram (ECG) showed supraventricular tachycardia (SVT) with a heart rate of 210 beats per minute and it was reverted to sinus rhythm with adenosine [Figure 1a]. Post-cardioversion ECG showed a counterclockwise loop, early RS transition, and high voltage complexes in all chest leads [Figure 1b].

Chest X-ray of the patient showed moderate cardiomegaly with dilated right atrium, dilated main pulmonary artery and right descending pulmonary artery with normal lung fields [Figure 2].

Transthoracic 2D echocardiography showed situs solitus, levocardia, single ventricle of left ventricular morphology and DILV, and a rudimentary right ventricle. The great arteries were malposed. Aorta was arising from LV and pulmonary artery from hypoplastic right ventricular chamber. Pulmonary artery was dilated with pulmonary stenosis with a peak gradient of 30 mm Hg. Left ventricular function was normal [Figure 3].

A cardiac magnetic resonance imaging (MRI) was done to confirm the diagnosis. It showed a DILV with large inlet ventricular septal defect (VSD), d-malposed great arteries, and dilated branch pulmonary artery with normal pulmonary and systemic venous drainage [Figure 4].

In view of SVT at presentation, an electrophysiological study was planned but deferred as the patient was not willing for further invasive interventions.

She was kept on oral medications with regular follow-ups.

DISCUSSION

DILV is a complex congenital heart defect characterized by the presence of two AV valve orifices leading into a single LV.[4] This condition results in a mixing of oxygenated and deoxygenated blood within the ventricle. While DILV is often associated with various other cardiac abnormalities, the coexistence of SVT is a rare occurrence and it adds an additional layer of complexity to diagnosis and management.

The exact mechanisms underlying SVT in DILV are not fully understood and are likely multifactorial. First, the abnormal anatomy of DILV, with the presence of dual AV valve orifices, may result in altered electrical conduction pathways and predispose the patient to arrhythmias. The abnormal positioning and arrangement of the AV valves can disrupt the normal electrical circuitry of the heart, leading to the development of SVT. Furthermore, the mixing of oxygenated and deoxygenated blood within the ventricle can cause hemodynamic disturbances, such as volume and pressure overload, which can trigger arrhythmias. In addition, the presence of associated cardiac abnormalities, such as VSDs or atrial septal defects,[5] can further contribute to the development of arrhythmias in DILV.

Diagnosing SVT in the context of DILV can be challenging due to the complexity of the underlying cardiac anatomy.

ECG is typically the initial diagnostic tool, but it may not always provide a clear distinction between SVT and other arrhythmias.

Further investigations, such as echocardiography, cardiac MRI, or electrophysiological studies, may be required to delineate the underlying anatomy and electrophysiological characteristics, aiding in accurate diagnosis and guiding management decisions.

Management of SVT in DILV requires an individualized approach based on the patient's clinical status and the severity of the arrhythmia. Initial management of

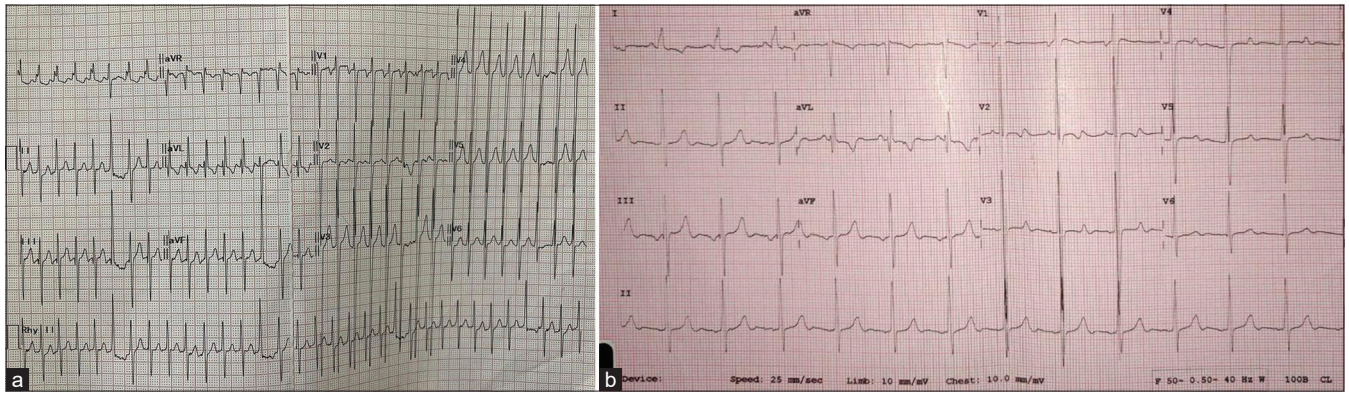

Figure 1: (a) Electrocardiogram (ECG) at presentation – Supraventricular tachycardia with a rate of 210/min. (b) ECG post-cardioversion – sinus rhythm, counter clockwise loop, early RS transition, and high voltage complexes.

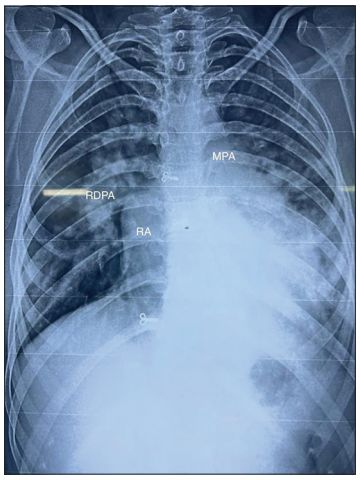

Figure 2: Chest X-ray of the patient showing moderate cardiomegaly with dilated RA; dilated MPA and RDPA: normal lung fields. RA: Right atrium, MPA: Main pulmonary artery, RDPA: Right descending pulmonary artery.

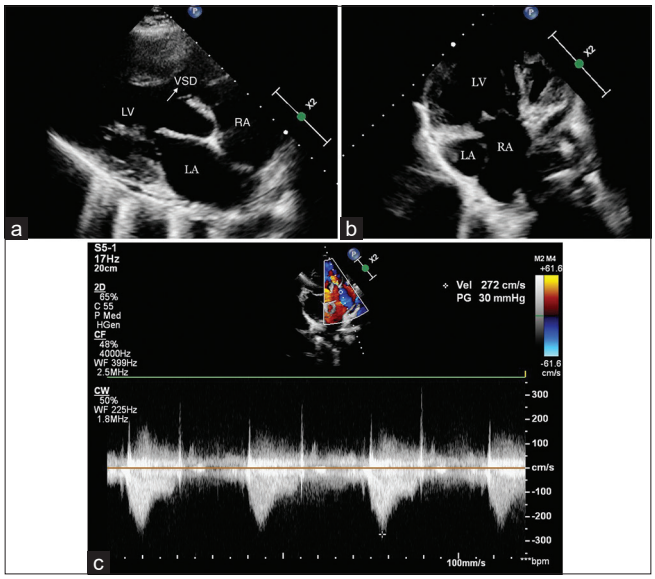

Figure 3: Transthoracic 2D echocardiography: (a) Conventional parasternal longitudinal axis view showing both LA and RA draining into LV with large inlet VSD. The chambers are labelled; LA: Left atrium, LV: Left ventricle, RA: Right atrium, VSD: Ventricular septal defect. (b) Modified parasternal longitudinal axis view showing both LA and RA draining into LV. The chambers are labelled; LA: Left atrium, LV: Left ventricle, RA: Right atrium. (c) Continuous wave Doppler showing peak gradient of 30 mm Hg across pulmonary valve.

termination of tachycardia depends on the hemodynamic status of the patient. Vagal maneuvers and pharmacological interventions (e.g., adenosine, beta-blockers, calcium channel blockers) can be used in stable patients.[6,7] In unstable cases, electrical cardioversion may be employed to restore sinus rhythm. In cases with recurrence, catheter-based interventions such as radiofrequency ablation may be considered.[6-8] Long-term management involves close monitoring for recurrence of SVT and addressing any underlying cardiac abnormalities that contribute to arrhythmogenesis. In certain cases, surgical interventions, such as AV valve repair or conversion to a univentricular circulation, may be necessary to optimize cardiac function and reduce the risk of arrhythmias.

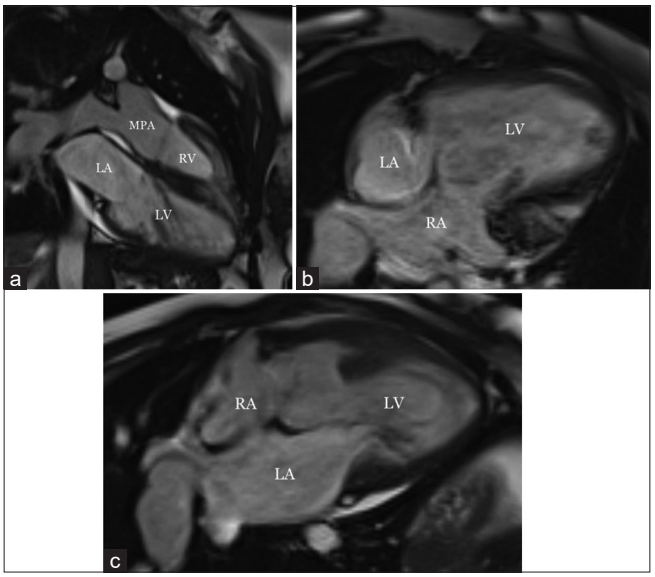

Figure 4: Cardiac magnetic resonance imaging: (a) Coronal view showing left atrium draining into left ventricle, and main pulmonary artery arising from rudimentary right ventricle. The chambers are labeled; LA: Left atrium, LV: Left ventricle, RA: Right atrium, RV: Right ventricle, MPA: Main pulmonary artery. (b and c) Axial view showing both LA and RA draining into LV. The chambers are labeled; LA: Left atrium, LV: Left ventricle, RA: Right atrium, RV: Right ventricle.

CONCLUSION

DILV is exceedingly unusual and survival without surgical intervention through the fourth decade of life is exceptional. The presentation of SVT in the setting of DILV is a rare occurrence that poses diagnostic and management challenges. This case of SVT unmasked a surprising culprit that defied conventional expectations, highlighting the need for comprehensive cardiac assessment in patients with arrhythmias.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Allen LD, Hornberger LK, Sharland GK. Double-Inlet Left Ventricle in the Fetus. London, United Kingdom: Greenwich Medical Media; 2000. p. 174-82.
2. Moodie DS, Ritter DG, Tajik AJ, O'Fallon WM. Long-term follow-up in the unoperated univentricular heart. Am J Cardiol 1984;53:1124-8.
3. Ammash NM, Warnes CA. Survival into adulthood of patients with unoperated single ventricle. Am J Cardiol 1996;77:542-4.
4. Perloff JK, Marelli AJ. Clinical Recognition of Congenital Heart Disease. 6th ed. Netherlands: Elsevier Inc.; 2012.
5. Bevilacqua M, Sanders SP, Van PS, Colan SD, Parness I. Double-inlet single left ventricle: Echocardiographic anatomy with emphasis on the morphology of the atrioventricular valves and ventricular septal defect. J Am Coll Cardiol 1991;18:559-68.
6. Khairy P, Van Hare GF, Balaji S, Berul CI, Cecchin F, Cohen MI, *et al*. PACES/HRS expert consensus statement on the recognition and management of arrhythmias in adult congenital heart disease: Developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Can J Cardiol 2014;30:e1-63.
7. Lundqvist CB, Potpara TS, Malmborg H. Supraventricular arrhythmias in patients with adult congenital heart disease. Arrhythm Electrophysiol Rev 2017;6:42-9.
8. Chiu SN, Wang JK, Lu CW, Wu KL, Tseng WC, Wu MH, *et al*. Electrophysiology study for complex supraventricular tachycardia in congenital heart disease patients with single-ventricle physiology. J Am Heart Assoc 2016;5:e004504.

How to cite this article: Syed I, Srinivas R, Balachander M, Nagula P. A tale of intricate connections: An interesting case of congenital heart disease with supraventricular tachycardia. Indian J Cardiovasc Dis Women. 2023;8:264-7.