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Single Ventricle in a 40-Year-Old Male Patient "Natural Evolution and Single Ventricle-Associated Complications in Adults: A Case Report"

Navarro Alonso Carlos, Gonzalez Sanchez David Emmanuel

Facultad de Medicina Unidad Saltillo, Universidad Autonoma de Coahuila, Saltillo, Coahuila, México
Email: medicinaus@uadec.edu.mx

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Abstract

Single ventricle is an uncommon congenital cardiac pathology with high mortality at early ages. However, due to new strategies and timely surgical treatment, it is increasingly seen in adults, which has increased the survival rate. This condition is characterized by a heart with a single functional ventricular cavity. The aim of this article is to report the case of a 40-year-old male with single ventricle, who underwent several surgeries in his childhood. After these surgical procedures, the patient lost medical follow-up and presented complications derived from the disease itself. Heart Failure is a common complication in people with single ventricle, and pharmacological heart failure therapies have been ineffective in mitigating the need of the heart transplantation. That's why it is essential to develop and apply new pharmacological techniques for the management of these patients in childhood as well as in adulthood. This would allow not only to avoid various comorbidities, but also to improve the quality of life of the patients.

Keywords

Single, Ventricle, Childhood, Survival, Comorbidities

1. Introduction

The term "single ventricle" has evolved from an anatomical definition, referring simply to a heart with a single ventricular cavity connected to the atria, to a description that encompasses both anatomical and functional aspects [1]. It now denotes a heart where both ventricles may be present with their full morphological characteristics, but one of them is hypoplastic and unable to perform its function

[1]. Consequently, hearts with a single ventricle can be classified into two categories: those with an atrioventricular (AV) connection in a single ventricle (either right or left) and those with a biventricular AV connection but a single outflow tract (either aortic or pulmonary) [1].

In hearts with an atrioventricular (AV) connection in a single ventricle and double inlet, several morphologies are possible depending on the cavity into which both AV valves terminate: left ventricle (63%), right ventricle (34%), or an undefined cavity (1%) [2] [3]. The prevalence of newborns with a univentricular heart is 3.1 to 4.9 per 100,000, while the prevalence in children living with this condition is 1.6 per 100,000, with much lower rates observed in adults [4].

Patients with a univentricular heart who undergo surgery in childhood and reach adulthood often develop comorbidities over time, including arrhythmias, coronary artery disease, cyanosis, heart failure, renal disease, and pulmonary problems, among others [3] [5]. Due to the rarity of this condition in adults and the complexity of the associated comorbidities, there is insufficient information to establish optimal medical treatment [4].

2. Clinical Case

2.1. Background of the Patient

A 40-year-old male patient with a history of double outlet right ventricle and pulmonary stenosis, diagnosed at 9 years of age, underwent a Blalock-Taussig systemic-pulmonary fistula to address his condition. This procedure was followed by an immediate postoperative complication due to obstruction, necessitating a subsequent intervention. The following year, the patient had cardiac catheterization and balloon angioplasty, which led to a modest improvement. However, a few months later, he required another surgery where a partial infundibulectomy was performed to resolve the pulmonary stenosis.

Subsequently, without further evaluations, the patient reported that throughout his life he had not experienced symptoms of heart failure, except for dyspnea classified as NYHA II. He noted that this dyspnea has worsened over the last 10 years, though it has not significantly affected his functional capacity. Additionally, the patient mentioned that his extremities have been cyanotic for the past 5 years.

2.2. Clinical Presentation

The patient presented to the emergency department with sudden-onset dyspnea classified as NYHA IV, accompanied by bendopnea and orthopnea. He reported that he was not engaged in any physical activity at the time. Upon admission, his vital signs were as follows: heart rate of 103 beats per minute, blood pressure of 90/60 mmHg, respiratory rate of 28 breaths per minute, and oxygen saturation of 76% on room air.

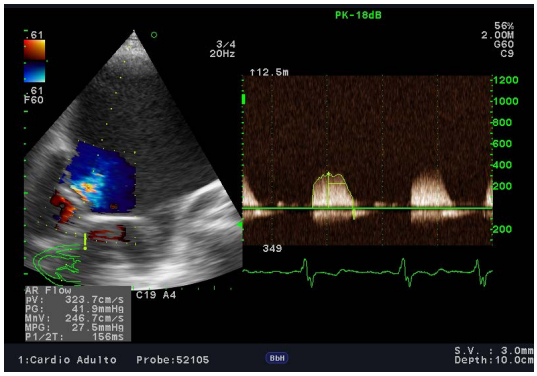
The patient stayed in the emergency department for 4 days. During this time, he required supplemental oxygen via a simple mask at 5 L/min, which improved his oxygen saturation to 92%. He received treatment for acute heart failure and

showed a favorable response. An EKG performed at admission (see Figure 2) revealed ventricular overload, pulmonary hypertension, atrial fibrillation, and right chamber enlargement.

Upon admission to the cardiology floor, a complete physical examination was performed. Clubbing was noted. Cardiac auscultation revealed localized holodiastolic murmurs at the aortic and pulmonary areas, and a holosystolic murmur at the tricuspid and mitral areas, graded 6/6, accompanied by an S3 heart sound. In the fifth intercostal space at the midclavicular line of the right hemithorax, a holosystolic murmur was heard with greater intensity compared to the left hemithorax, along with high-intensity fremitus. No carotid murmurs were detected, and grade I jugular ingurgitation was observed on inspection.

2.3. Complementary Studies and Imagine Test

Transthoracic Echo: Reported with the presence of single double outlet ventricle with a Left Ventricular Eject Fraction of 20%, together with the presence of severe aortic insufficiency, double pulmonary lesion with stenosis and mild pulmonary insufficiency, type 1 diastolic dysfunction, non-dilated LA and RA, moderate mitral and tricuspid insufficiency, without the presence of pericardial effusion (Figure 1).



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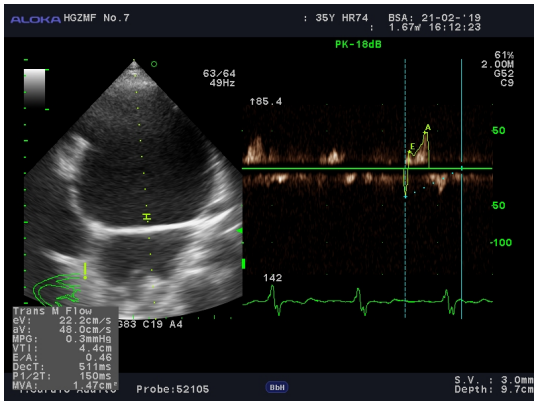
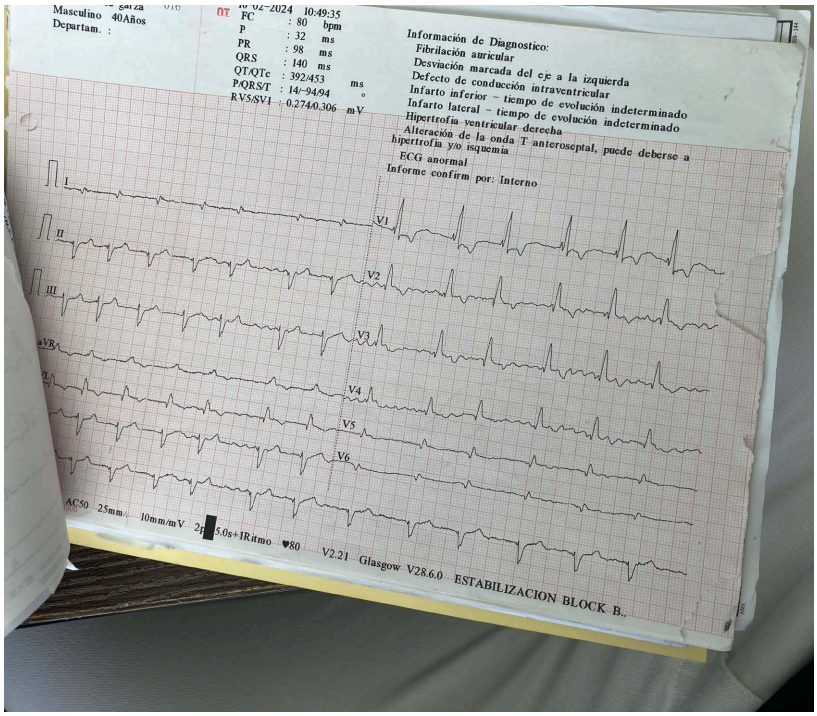


(b)

Figure 1. Single ventricle is shown together with the presence of left and right atrium. A-Se demonstrates regurgitant jet pattern at the level of the left atrium. of aortic valve.

Electrocardiogram: HR 80 lpm, axis deviated to the left (−90˚), presence of Atrial Fibrilation with intermediate ventricular response and presence of complete BRDHH, HVD by Cabrera index (0.86) and ventricular systolic overload (Figure 2).



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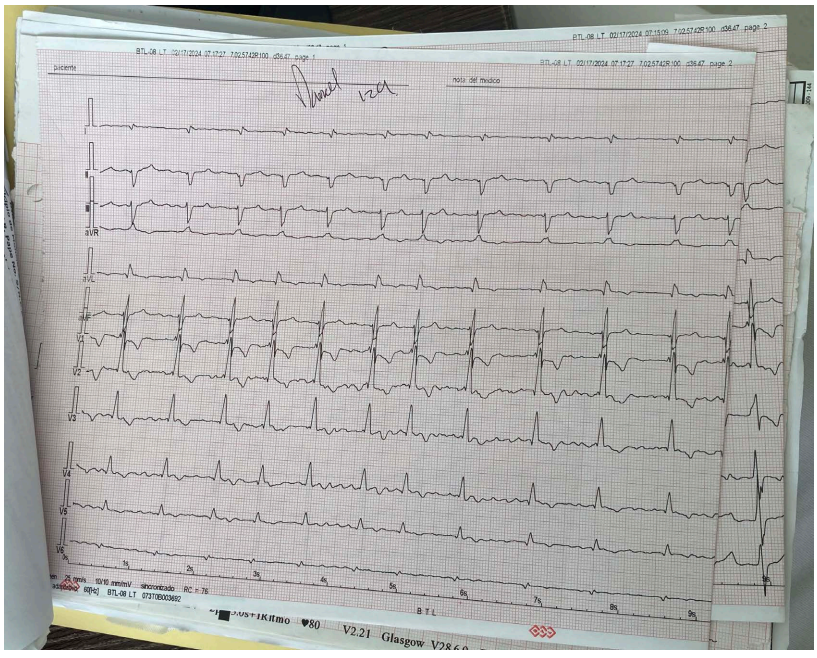

(b)

Figure 2. EKG on admission to the emergency department (a) and during the stay on the cardiology floor (b).

Chest X-ray PA: Presence of Cardiomegaly grade IV with cardiothoracic index of 0.68 (Figure 3).

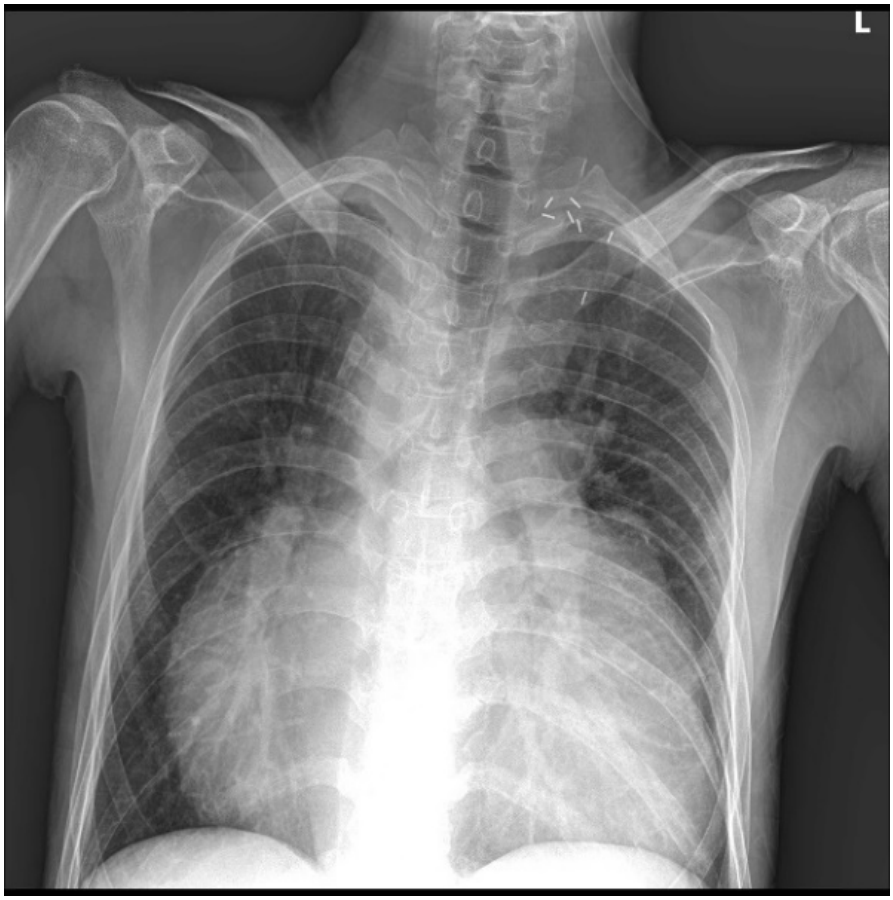

Figure 3. Chest X-Ray with cardiomegaly grade IV and increase vascular marking.

Laboratory studies (Table 1) and (Table 2).

Table 1. Laboratory studies and reference values.

TEST NAME	BLOOD CELLS COUNT AND CLOTTING TIME		
	RESULT	UNITS	REFERENCE RANGE
LEUKOCYTES	4.4	10 × 3/Ml	4.5 - 11.0
HEMOGLOBIN	20.3	g/dL	14.00 - 18.00
HEMATOCRIT	60.6	%	42 - 52
PLATELETS	130	X10 × 3	150.00 - 450.00
MCV	96.74	Fl	83 - 100
HCM	32.4	Pg	28 - 32
PT	13.6	Seg	12 - 14
TTP	41.9	Seg	28 - 36
INR	1.19		0.9 - 1.5

Table 2. Laboratory studies and reference values.

TEST NAME	BLOOD CHEMISTRY AND LIVER FUNCTION TEST		
	RESULT	UNITS	REFERENCE RANGE
GLUCOSE	62	mg/dl	70 - 105
UREA	64	mg/dl	10 - 50
CREATININE	0.99	mg/dl	0.6 - 1.3
BUN	30	mEq/L	135 - 145
NA	135	mEq/L	3.5 - 5.3
K	4.6	mEq/L	96 - 109
CL	107	U/ml	24 - 194
CPK	81	mg/dl	140 - 200
CHOLESTERO	149	mg/dl	0.32 - 1.08
BT	3.8	mg/dl	0.08 - 0.72
BI	2.7	mg/dl	0.10 - 0.50
BD	1.1	g/dL	3.5 - 5.5
ALBUMIN	4.2	mg/Dl	3.4 - 7.0
URIC ACID	8.0	U/L	8 - 40
TGO	33	U/L	10 - 40
TGP	29	U/L	44 - 147
FA	97	U/L	125 - 220
DHL	238	U/L	140 - 280

2.4. Treatment

The patient is currently receiving treatment with Sildenafil 50 mg OV c/24hrs (due to the double pulmonary lesion with stenosis and type I diastolic dysfunction), Spironolactone 25 mg OV c/24hrs (as part of heart failure treatment), ASA 150 mg OV c/24hrs, Rivaroxaban 20 mg OV c/24hrs and Metoprolol 25 mg OV c/24hrs. Treatment was also given with sacubitril-valsartan 50 mg for two single doses because the patient presented hypotension after its administration, so it was decided to suspend it. Also during his stay on the cardiology floor he underwent phlebotomy due to the presence of polyglobulia secondary to congenital heart disease. Currently, due to clinical improvement and adequate response to treatment, he does not require supplemental O2 support.

3. Discussion

Single ventricle is an extremely rare congenital heart defect in newborns and even less common in adults, largely due to the numerous comorbidities associated with the condition, which often lead to early mortality [5]. When discussing single ventricle physiology, it can be classified based on which ventricle receives the two atrioventricular (AV) valves, with the left ventricle being the most common

recipient (63% - 70%) [2] [6]. However, in the patient presented in this case, transthoracic echocardiogram (TTE) and chest X-ray findings, along with EKG data, confirm that both AV valves lead to the right ventricle.

Patients with single ventricle physiology frequently experience heart failure at diagnosis and progressive heart failure over time, which is the most common cause of mortality in these individuals [4]. They may also have extracardiac comorbidities such as thyroid dysfunction, renal disease, liver cirrhosis, and arrhythmias (typically atrial tachyarrhythmias), which contribute to their overall morbidity [5].

The clinical presentation of these patients depends on the presence or absence of large vessel flow, resistances, and the morphology and function of the predominant ventricle [1]. For this patient, the clinical presentation includes congestive heart failure due to ventricular volume overload secondary to pulmonary insufficiency and insufficiency of both AV valves.

In pediatric patients, various palliative surgical procedures are available depending on the specific physiology [6] [7]. In adults, particularly those who are not candidates for Fontan surgery (which involves creating an atriopulmonary shunt and managing Cavo pulmonary flow), other palliative surgical options are considered [8]. If these interventions are not feasible, cardiac transplantation may be pursued to improve survival and quality of life. However, the feasibility of transplantation depends on the presence of pulmonary stenosis, which can protect the pulmonary circulation from hyperflow. If pulmonary stenosis is not present, cardiopulmonary transplantation may be recommended [8].

4. Conclusion

In conclusion, while advancements have improved survival and quality of life for patients with single ventricle physiology, continuous and careful follow-up throughout their lives is essential [9]. This includes regular paraclinical and imaging studies, such as transthoracic echocardiograms, to promptly address all associated comorbidities [7] [10]. Given that treatment in childhood often relies on surgical interventions, it is crucial to develop and implement new pharmacological strategies for the management of these patients from childhood through adulthood [4] [5]. Such strategies could help mitigate various comorbidities and provide pharmacological options when surgical interventions are not feasible. Additionally, a multidisciplinary approach is vital due to the complex nature of the associated pathologies.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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