Cardiovascular and Metabolic Science www.ancam.org.mx

Clinical case

Vol. 35 No. 1
January-March 2024

doi: 10.35366/115000

Complete heart block in an adult patient with isolated congenitally corrected transposition of the great arteries and *situs inversus*

Bloqueo auriculoventricular completo en un paciente adulto con transposición congénitamente corregida de grandes arterias aisladas y situs inversus

José Martín Alanís-Naranjo,*,‡ Cielmar Galeana-Abarca,*,§ Virginia Campos-Garcilazo*,¶

Keywords:
congenitally corrected transposition of the great arteries, complete heart block, congenital heart disease, adult, *situs inversus*.

Palabras clave:
transposición congénitamente corregida de grandes arterias, bloqueo auriculoventricular completo, cardiopatía congénita, adulto, situs inversus.

ABSTRACT

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart disease. Many patients remain asymptomatic when no cardiac lesions are present (isolated ccTGA). In ccTGA, the atrioventricular conduction system may be abnormal, resulting in progressive dysfunction and, eventually, a Complete Heart Block (CHB). In ccTGA with *situs inversus*, the conduction pathway resembles a normal tract, but the atrioventricular node is located posteriorly. Compared with *situs solitus*, spontaneous CHB is uncommon in ccTGA patients with *situs inversus*. We report the case of a 40-year-old female without previous medical conditions or having a family or personal history of heart disease presented with loss of consciousness. At admission, electrocardiography revealed bradycardia, CHB, and hypertrophy of the right ventricle. Cardiovascular imaging tests detected an isolated ccTGA with *situs inversus* and levocardia. The Holter monitor revealed intermittent CHB. Exercise testing demonstrated chronotropic incompetence. An epicardial pacemaker was implanted, and the patient was discharged symptom-free. At a two-years follow-up remains asymptomatic. This case illustrates the importance of cardiovascular imaging in defining cardiac anatomy, ruling out other congenital heart defects, and facilitating pacing therapy in complex congenital heart disease. Congenital heart disease patients should be treated by a multidisciplinary team with expertise in permanent pacing.

RESUMEN

La transposición congénitamente corregida de grandes arterias (TccGA) es una cardiopatía congénita poco común. Muchos pacientes permanecen asintomáticos cuando no hay lesiones cardiacas presentes (TccGA aislada). En la TccGA, el sistema de conducción auriculoventricular puede ser anormal, lo que resulta en una disfunción progresiva y, finalmente, en un bloqueo auriculoventricular (BAV) completo. En la TccGA con situs inversus, la vía de conducción se asemeja a un tracto normal, pero el nódulo auriculoventricular se ubica posteriormente. En comparación con el situs solitus, el BAV completo espontáneo es poco común en pacientes con TccGA con situs inversus. Se presenta el caso de una mujer de 40 años sin enfermedades previas ni con antecedentes familiares o personales de cardiopatías que presentó pérdida de conciencia. Al ingreso, el electrocardiograma reveló bradicardia, BAV completo e hipertrofia del ventrículo derecho. Estudios de imagen cardiaca detectaron una TccGA aislada con situs inversus y levocardia. El monitoreo Holter reveló BAV completo intermitente. Las pruebas de esfuerzo demostraron incompetencia cronotrópica. Se le implantó un marcapasos epicárdico y la paciente fue dada de alta asintomática. Tras dos años de seguimiento permaneció asintomática. Este caso ilustra la importancia de imagen cardiovascular para definir la anatomía cardiaca, descartar otras cardiopatías congénitas y facilitar la terapia de estimulación cardiaca en cardiopatías congénitas complejas. Los pacientes con cardiopatías congénitas deben ser tratados por un equipo multidisciplinario con experiencia en estimulación cardiaca permanente.

* Hospital Regional 1º de Octubre, ISSSTE, Mexico City, Mexico.
‡ Internist, Cardiology Resident. ORCID: 0000-0001-6631-7228
§ Cardiologist, Cardiovascular Imaging Specialist.
¶ Cardiologist, Cardiac Electrophysiologist.

Received:
11/21/2023
Accepted:
02/21/2024

How to cite: Alanís-Naranjo JM, Galeana-Abarca C, Campos-Garcilazo V. Complete heart block in an adult patient with isolated congenitally corrected transposition of the great arteries and *situs inversus*. Cardiovasc Metab Sci. 2024; 35 (1): 16-21. https://dx.doi.org/10.35366/115000

INTRODUCTION

Acongenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart disease characterized by discordant atrioventricular and ventriculo-arterial connections.[1,2] The right atrium (RA) enters the morphological left ventricle (LV), which rises to the pulmonary artery, and the left atrium (LA) communicates with the morphological right ventricle (RV), which gives rise to the aorta.[2] Among the associated lesions are ventricular septal defects (70%), pulmonary stenosis (40%), and dysplastic systemic tricuspid valves.[1] The clinical course of adults with ccTGA is relatively unknown, with cases of isolated ccTGA rarely developing complications before adulthood.[1,3,4] In most cases, the position of the atrioventricular node and the bundle of his is abnormal, resulting in abnormal atrioventricular conduction.[1] In patients with ccTGA and *situs inversus*, complications are lower than in patients with situs solis, including Complete Heart Block (CHB).[4] Visceroatrial *situs inversus* with ccTGA is a rare congenital condition.[5,6] Given the rarity of this condition, there are limited reports of CHB in adult patients with isolated ccTGA and *situs inversus*. Therefore, we report a case of symptomatic intermittent CHB in an adult patient with ccTGA, *situs inversus*, and levocardia who required pacing therapy.

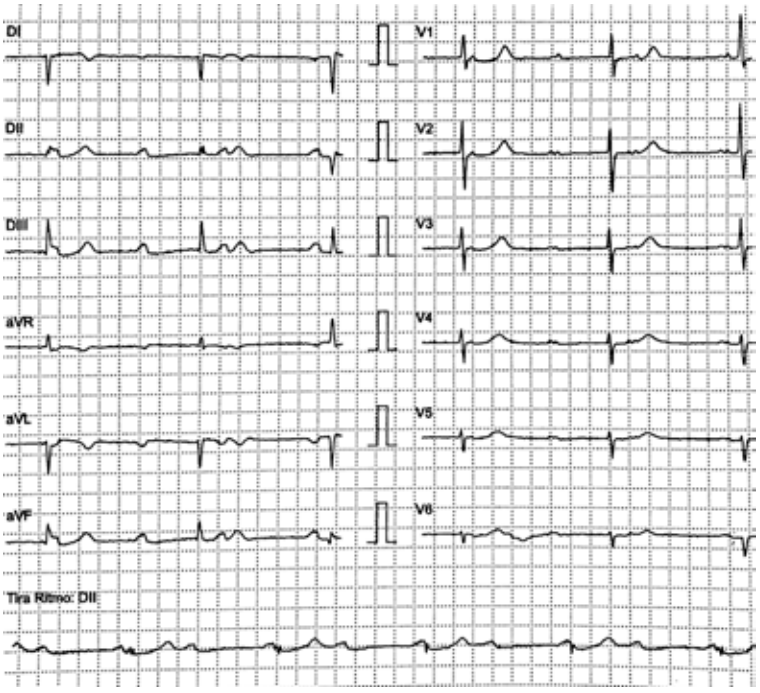

Figure 1: Initial 12-Lead electrocardiogram: atrioventricular dissociation, right ventricular hypertrophy, normal QRS voltage and duration, a prolonged QT interval, no ST-segment elevation or depression, and negative T waves in leads DI, aVR, and aVL.

CASE PRESENTATION

A 40-year-old female was admitted to the emergency department after briefly losing consciousness at work in the last 12 hours. The patient felt well before and immediately following the episode, and there was no apparent trauma or confusion. As part of her initial evaluation at another center, an electrocardiogram revealed a CHB *(Figure 1)*, which led to her referral to this facility. The patient denied having any previous medical conditions or having a family or personal history of heart disease and also denied taking any medications.

On examination at admission, she was afebrile, with a blood pressure of 125/67 mmHg and a heart rate of 40 beats/min. The cardiovascular examination revealed a high-pitched and loud grade 4/6 pansystolic murmur in the tricuspid valve area, which increased upon inspiration.

On admission, electrocardiography revealed a CHB with right ventricular hypertrophy *(Figure 2)*. In addition, the blood tests were relevant for high cholesterol and triglyceride levels, with the rest of the studies in the normal range *(Table 1)*.

Transesogapheal echocardiography revealed a mirror-image atrial arrangement, with right-sided LA connected to a morphological RV with an emerging aorta, a left-sided RA connected to a morphologic LV with an arising pulmonary artery, a right ventricular ejection fraction (RVEF) of 38%, an estimated pulmonary artery systolic pressure of 106 mmHg, mild pulmonary regurgitation, intact interatrial and interventricular septums, moderate tricuspid regurgitation, and no intracavitary thrombus. Cardiac computed tomography revealed an isolated ccTGA with visceroatrial *situs inversus* and levocardia *(Figures 3 and 4)*.

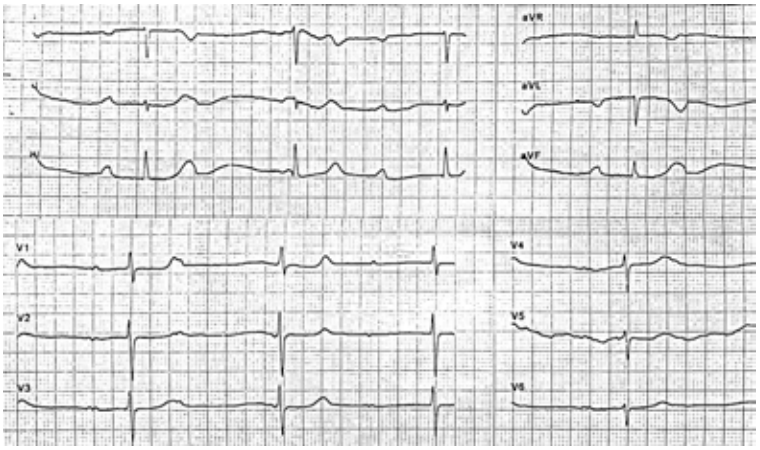

Figure 2: 12-Lead electrocardiogram on admission: atrioventricular dissociation, right ventricular hypertrophy, normal QRS voltage and duration, a prolonged QT interval, no ST-segment elevation or depression, and negative T waves in leads DI, aVR, and aVL.

The Holter monitor showed intermittent CHB alternated with sinus rhythm and premature ventricular contractions *(Figure 5)*. In the exercise testing, the patient achieved 4.6 METS, showed chronotropic incompetence, and was suspended by fatigue *(Figure 6)*.

Based on the results of these studies, a permanent epicardial pacemaker was implanted; the patient was discharged symptom-free and with a functioning pacemaker. At a two-year follow-up, the patient remained asymptomatic, and a TTE showed RVEF 38% with moderate tricuspid regurgitation with a functional pacemaker.

DISCUSSION

Described by Von Rokitansky in 1875, ccTGA accounts for approximately 0.5% of all congenital heart defects, with limited information on adult patients.[7]

Concerning the conduction system, the atrioventricular node and His bundle follow an unusual path, and many patients have dual AV nodes. Second anomalous AV nodes and bundles are usually anterior, and the long, penetrating bundle is prone to fibrosis with increasing age. As a result, the conduction system is somewhat tenuous, with the

Table 1: Blood tests on admission.

Test	Result	Reference [range]
Leukocytes (10^3/μL)	7.02	5-10
Hemoglobin (g/dL)	14.4	14-18
Hematocrit (%)	45.7	42-52
MCV (fL)	78.9	80-95
MCH (pg)	24.9	27-31
MCHC (g/dL)	31.7	32-36
Platelets (10^3/μL)	280	130-400
Glucose (mg/dL)	103	74-106
Creatinine (mg/dL)	1.02	0.6-1.2
Urea (mg/dL)	30.3	18-50
AST (U/L)	39	13-39
ALT (U/L)	52	7-52
Total bilirrubin (mg/dL)	0.56	0.3-1
Albumin (g/dL)	3.9	3.5-5.7
Globulin (g/dL)	2.9	1.9-2.7
Total protein (g/dL)	6.8	6.4-8.9
LDH (U/L)	195	140-271
GGT (U/L)	60	9-64
AP (U/L)	114	34-104
Creatin kinase (U/L)	83	21-232
Creatin kinase MB (U/L)	13.8	0-10
Troponin I (ng/L)	< 1.5	0-19
Sodium (mEq/L)	136	136-145
Potassium (mEq/L)	4.2	3.5-5.1
Chloride (mEq/L)	106	98-107
Total cholesterol (mg/dL)	223	0-200
Triglyceride (mg/dL)	232	0-150
HDL (mg/dL)	46	32-92
HbA1c (%)	5.7	4-6.5
Total T3 (ng/mL)	0.87	0.87-1.78
Total T4 (μg/dL)	8.41	6.09-12.23
Free T4 (ng/dL)	0.83	0.61-1.12
TSH (UI/mL)	1.73	0.34-5.6

MCV = mean corpuscular volume. MCH = mean corpuscular hemoglobin. MCHC = mean corpuscular hemoglobin concentration. AST = aspartate aminotransferase. ALT = alanine aminotransferase. LDH = lactate dehydrogenase. GGT = gamma glutamyltransferase. AP = alkaline phosphatase. HDL = high-density lipoprotein. HbA1c = glycosylated hemoglobin. T3 = triiodothyronine. T4 = thyroxine. TSH = thyroid-stimulating hormone.

incidence of complete AV block rising at 2% per year.[2,8,9]

In 10% of ccTGA cases, *situs inversus* is present; in this condition, the conduction pathway resembles a normal tract with an atrioventricular node located posteriorly.[4,10] Levocardia usually exists with *situs solitus*, but dextrocardia can complicate the anatomical presentation in approximately 25% of the cases.[10]

In the literature, no significant differences in ventricular septal defect, pulmonary tract stenosis, and long-term mortality were found between ccTGA with *situs inversus* or *situs solitus*. There was, however, a lower incidence of nonsurgically related CHB (0 vs 42%, p = 0.032), development of CHB in 7.4 years (12.5 vs 57%, p = 0.045), tricuspid valve anomalies (0.0 vs 50%, p = 0.01), overall complications (25 vs 73%, p = 0.034) and heart failure (p = 0.038).[4]

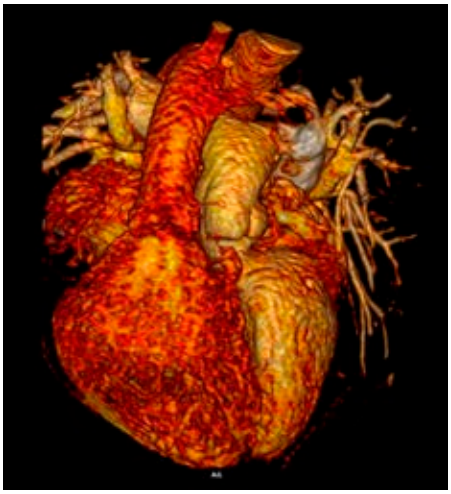

Figure 4: Cardiac computed tomography: 3D reconstruction exhibiting right-sided anterior aorta emerging from right ventricle with a left-sided posterior pulmonary trunk arising from left ventricle.
RV = right ventricle. LV = left ventricle.

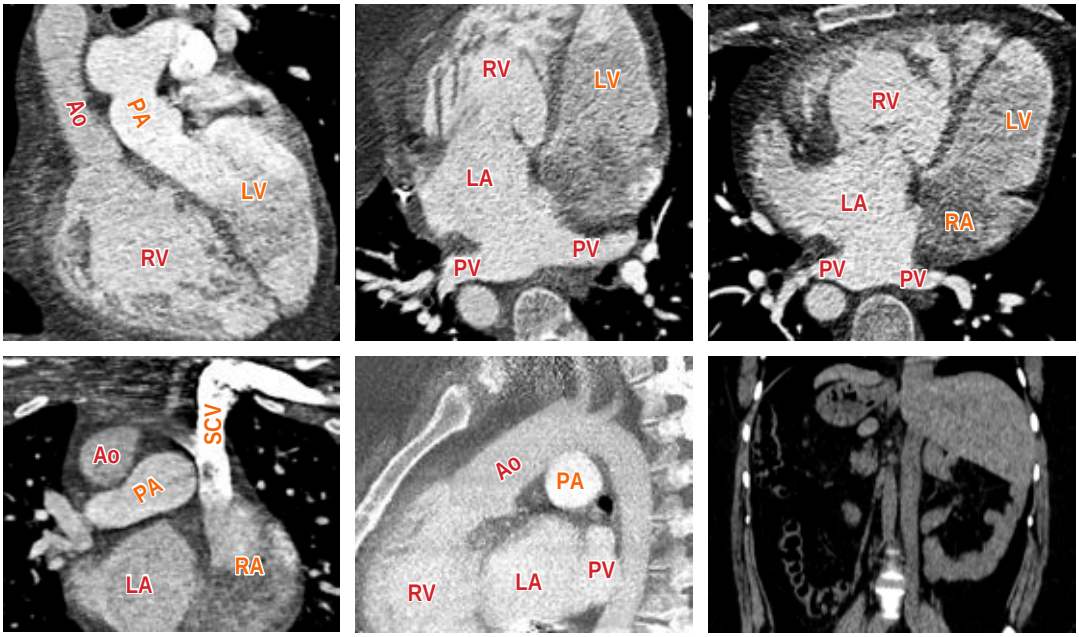

Figure 3: Cardiac computed tomography: visceroatrial *situs inversus*: left-sided inferior cava vein, right-sided abdominal aorta, right-sided liver, left-sided stomach, mirror-image atrial arrangement with L-looping of ventricles. Ventriculoarterial discordance: a right-sided morphologic right ventricle with an arising aorta and a left-sided morphologic left ventricle with an emerging pulmonary artery. Atrioventricular discordance: right-sided left atrium receiving pulmonary vein drainage communicates with the tricuspid valve to right ventricle, while left-sided right atrium receiving cava vein drainage communicates with the mitral valve to left ventricle.
RV = right ventricle. LV = left ventricle. LA = left atrium. RA = right atrium. Ao = aortic. PA = pulmonary artery. PV = pulmonary vein. SCV = superior cava vein.

The radiographic and electrocardiographic findings often lead to the diagnosis of ccTGA when symptoms are absent and physical findings are subtle.[1,8,10] Electrocardiograms frequently reveal ventricular hypertrophy and atrial enlargement, even in the absence of other cardiac lesions.[1,10] To confirm the diagnosis of ccTGA, cardiac imaging is essential, which aids in defining cardiac anatomy despite clinical suspicion.[1]

Pacemaker implantation may be justified in patients who suffer from symptomatic chronotropic incompetence. Current recommendations for cardiac pacing in patients with congenital heart disease and high-degree atrioventricular block are as follows: a)

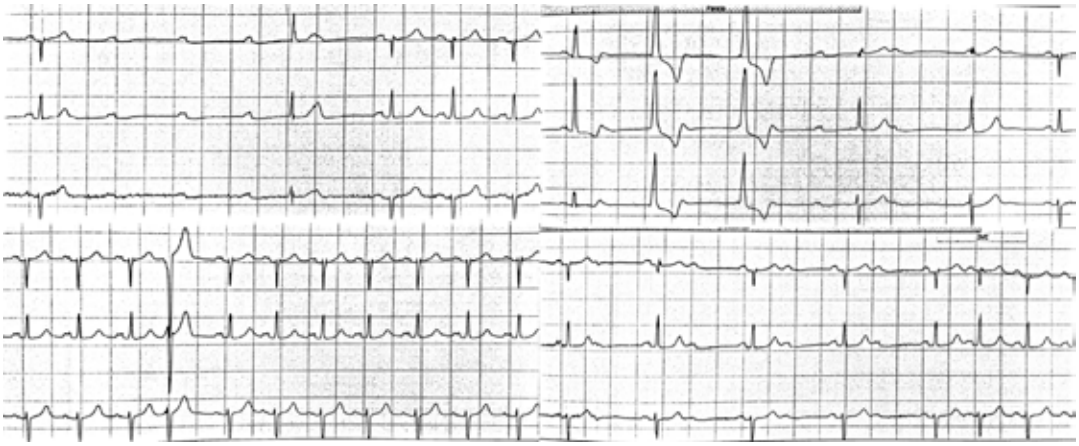

Figure 5: Holter monitor: intermittent complete heart block alternated with sinus rhythm and premature ventricular contractions.

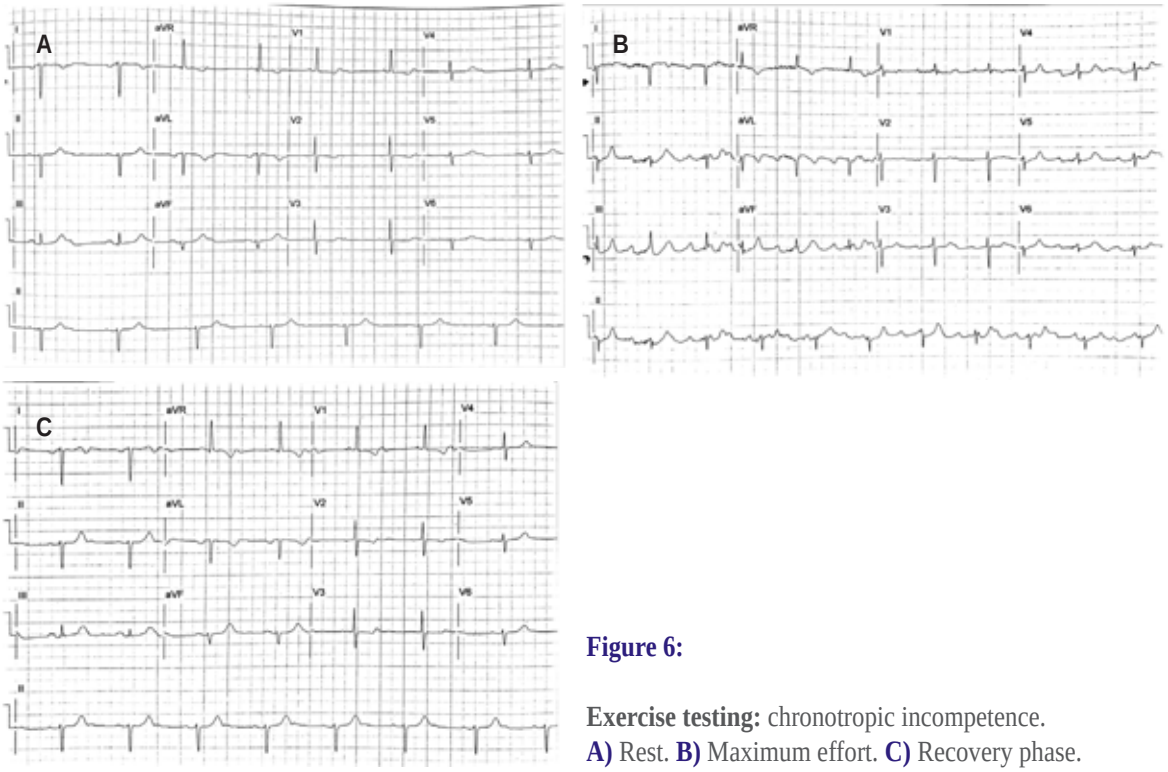

Figure 6:

Exercise testing: chronotropic incompetence. **A)** Rest. **B)** Maximum effort. **C)** Recovery phase.

symptoms, b) pauses > 3 times the cycle length of the ventricular escape rhythm, c) broad QRS escape rhythms, d) prolonged QT interval, e) complex ventricular ectopy, and f) the mean daytime heart rate < 50 beats/min.[11]

The patients have a reduced life expectancy; 50% would still be alive at 60 years of age if they did not have associated lesions. Additionally, no evidence exists that medical treatment is associated with preventing heart failure or improving outcomes.[1]

Surgery is generally reserved for symptomatic and asymptomatic patients with evidence of deteriorating RV function and worsening tricuspid regurgitation. Intervention in asymptomatic patients with preserved RV and tricuspid valve functions without any other septal defects is controversial.[10]

CONCLUSIONS

Isolated ccTGA usually does not manifest symptoms until adulthood; CHB results from abnormalities in the atrioventricular conduction system associated with discordant atrioventricular and ventriculoarterial connections. Patients with *situs inversus* and ccTGA have a lower incidence of CHB than those with *situs solitus*. In this group of patients, cardiovascular imaging studies are essential for defining cardiac anatomy and facilitating pacing therapy.

REFERENCES

1. Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP et al. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021; 42(6): 563-645.
2. Warnes CA. Transposition of the great arteries. Circulation. 2006; 114(24): 2699-2709.
3. Auer J, Pujol C, Maurer SJ, Nagdyman N, Ewert P, Tutarel O. Congenitally corrected transposition of the great arteries in adults-a contemporary single center experience. J Cardiovasc Dev Dis. 2021; 8(9): 113.
4. Oliver JM, Gallego P, Gonzalez AE, Sanchez-Recalde A, Brett M, Polo L et al. Comparison of outcomes in adults with congenitally corrected transposition with *situs inversus* versus *situs solitus*. Am J Cardiol. 2012; 110(11): 1687-1691.
5. Sohns JM, Steinmetz M, Schneider H, Fasshauer M, Staab W, Kowallick JT et al. *Situs inversus* totalis with congenitally corrected transposition of the great arteries: insights from cardiac MRI. Springerplus. 2014; 3: 601.
6. Lachhab F, Ajaja R, Fikri I, Amellal M, Otmani Y, Amri R et al. Transposition of the great arteries in *situs inversus totalis*. World J Pediatr Congenit Heart Surg. 2011; 2(3): 520-522.
7. Amaral F, Valente AM, Manso PH, Gali LG, Braggion-Santos MF, Rocha JM et al. Congenitally Corrected Transposition of the Great Arteries in the Adult. Braz J Cardiovasc Surg. 2022; 37(4): 534-545.
8. Kutty S, Danford DA, Diller GP, Tutarel O. Contemporary management and outcomes in congenitally corrected transposition of the great arteries. Heart. 2018; 104(14): 1148-1155.
9. Spigel Z, Binsalamah ZM, Caldarone C. Congenitally corrected transposition of the great arteries: anatomic, physiologic repair, and palliation. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2019; 22: 32-42.
10. Kumar TKS. Congenitally corrected transposition of the great arteries. J Thorac Dis. 2020; 12(3): 1213-1218.
11. Glikson M, Nielsen JC, Kronborg MB, Michowitz Y, Auricchio A, Barbash IM et al. 2021 ESC Guidelines on cardiac pacing and cardiac resynchronization therapy. Eur Heart J. 2021; 42(35): 3427-3520.

Declaration of patient consent: the authors declare they have followed their workplace protocols for using patient data. Also, they certify that the patient has received sufficient information and has given written informed consent for her images and other clinical information to be reported in the journal, without names or initials, to protect the right to privacy.

Funding: no financial support was received for this study.

Conflict of interest: the authors declare no conflict of interest.

Correspondence:
Virginia Campos-Garcilazo
E-mail: vickycampos2000@yahoo.com