

Case report

Double-Inlet single ventricle associated with transposition of the great arteries, *situs inversus*, and criss-cross heart in an unrepaired adult: A case report

Brayan Vargas-Lucio^{1,a}, Jaime Bravo-Alvarado^{2,b}, Sebastián Gaibor-Gaibor^{3,a}, Erick Vargas-Lucio^{1,a}, Alex Almache-Palacios^{1,a}

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Author affiliations

¹ Escuela Superior Politécnica de Chimborazo, Riobamba, Ecuador.
² Hospital General Puyo, Puyo, Ecuador.
³ Universidad de las Américas, Quito, Ecuador.
^a General physician.
^b Cardiologist.

Correspondence

Brayan José Vargas Lucio
Escuela Superior Politécnica de Chimborazo, Panamericana Sur km 1 ½ entre Av. Canónigo Ramos y Av. 11 de Noviembre, código postal EC060155, Riobamba, Ecuador.

Email

vargas.brayanj@gmail.com

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ABSTRACT

The single ventricle represents a group of complex congenital heart diseases characterized by the mixing of oxygenated and deoxygenated blood, chronic cyanosis, and ventricular volume overload. The combination of a double-inlet single ventricle with transposition of the great arteries, *situs inversus*, and a criss-cross heart is extremely rare, reported only in isolated cases in the literature. Most of these patients require early palliative interventions, such as the bidirectional Glenn procedure and eventual Fontan circulation, to optimize oxygenation and reduce ventricular overload. We present the case of a 26-year-old man with a history of cyanotic congenital heart disease, scheduled for surgery at age 12 but managed conservatively due to high surgical risk. Survival into adulthood without definitive surgical correction is likely due to a favorable hemodynamic balance conferred by pulmonary stenosis, which limits pulmonary overcirculation and protects the dominant ventricle. Currently, the patient remains clinically stable, classified as New York Heart Association functional class II, with no recent episodes of cardiovascular decompensation, hospitalizations, or documented thromboembolic events. This case highlights the anatomical rarity and underscores the importance of specialized follow-up in adults with complex univentricular congenital heart disease who have not undergone corrective surgery.

Keywords: Univentricular Heart; Transposition of Great Vessels; Situs Inversus; Heart Defects, Congenital (Source: MeSH-NLM).

RESUMEN

Ventrículo único de doble entrada asociado a transposición de grandes arterias, *situs inversus* y corazón *criss-cross* en adulto no corregido: reporte de caso

El ventrículo único constituye un grupo de cardiopatías congénitas complejas caracterizadas por la mezcla de sangre oxigenada y no oxigenada, cianosis crónica y sobrecarga de volumen ventricular. La combinación de ventrículo único de doble entrada con transposición de las grandes arterias, *situs inversus* y corazón tipo *criss-cross* es extremadamente rara, reportada solo en casos aislados en la literatura. La mayoría de estos pacientes requieren intervenciones paliativas tempranas, como la derivación bidireccional de Glenn y la eventual circulación de Fontan, para optimizar la oxigenación y reducir la sobrecarga ventricular. Presentamos el caso de un hombre de 26 años con antecedente de cardiopatía congénita cianógena, programado para cirugía a los 12 años, pero manejado de forma conservadora debido al alto riesgo quirúrgico y decisión familiar. La supervivencia hasta la adultez sin corrección quirúrgica probablemente se debe al equilibrio hemodinámico favorecido por la estenosis pulmonar, que limita la sobrecirculación pulmonar y protege al ventrículo dominante. Actualmente, el paciente se mantiene clínicamente estable, en clase funcional II de la New York Heart Association, sin episodios recientes de descompensación cardiovascular, hospitalizaciones ni eventos tromboembólicos documentados. Este caso resalta la rareza anatómica y la importancia del seguimiento especializado en adultos con cardiopatías univentriculares complejas no intervenidas.

Palabras clave: Corazón Univentricular; Transposición de los Grandes Vasos; Situs Inversus; Cardiopatías Congénitas (Fuente: DeCS-BIREME).

Introduction

Single-ventricle (SV) physiology encompasses a group of congenital heart defects (CHDs) that are not amenable to conventional biventricular repair. These defects have an estimated prevalence of 4-8 per 10,000 live births and account for approximately 7.7% of CHDs diagnosed in childhood ⁽¹⁾. A single ventricle with multiple associated anomalies represents a highly heterogeneous spectrum of CHDs. Criss-cross heart itself is extremely rare, with an incidence of 8 cases per 1,000,000 live births, accounting for less than 0.1% of all CHDs described in recent series ⁽²⁾. Moreover, the coexistence of *situs inversus totalis* with complex cardiac malformations, such as transposition of the great arteries (TGA), has been documented only in isolated case reports, reflecting its rarity and limited representation in contemporary series ⁽³⁾.

SV physiology is characterised by mixing of oxygenated and deoxygenated blood, volume overload, and haemodynamics governed by the relative pulmonary and systemic vascular resistances ⁽⁴⁾. Staged cavopulmonary palliation, comprising a bidirectional Glenn procedure followed by completion of the Fontan circulation, is the predominant strategy for improving oxygenation and reducing ventricular workload ^(4,5). In the largest reported cohort, the EuroFontan Registry, which included more than 3,500 patients, the median age at Fontan completion was 4.2 years, with early mortality below 1% and a composite adverse outcome rate of approximately 5% ⁽⁶⁾. Despite advances in surgical and medical management, some patients reach adolescence or adulthood without definitive surgery, with persistently suboptimal oxygen saturation and functional limitation; such cases are exceptional. We describe a young adult with a double-inlet single ventricle who was referred for surgery at the age of 12 years but received pharmacological management, and in whom clinical follow-up and supportive care have maintained functional and haemodynamic stability.

Case report

A 26-year-old man living in a tropical region of Ecuador, at an altitude of 950 m above sea level, had a history of cyanotic CHD diagnosed in the neonatal period. Surgical correction was scheduled at the age of 12 years but was not performed because his family declined the intervention after the treating medical team explained the associated procedural risks.

He was referred from primary care for cardiological assessment and follow-up of complex CHD with single-ventricle physiology. He reported mild exercise intolerance, with dyspnoea and fatigue during moderate physical activity. Physical examination showed longstanding peripheral cyanosis and digital clubbing (**Figure 1**). Cardiac auscultation revealed a grade VI/VI systolic ejection murmur over the pulmonary area, accompanied by a palpable thrill. Resting peripheral oxygen saturation was 86% on room air. Given that

symptoms occurred only during ordinary physical activity and were absent at rest, he was classified as New York Heart Association (NYHA) functional class II.

Laboratory investigations showed a haemoglobin concentration of 20.9 g/dL and a haematocrit of 60.5%, consistent with secondary erythrocytosis, for which he underwent therapeutic phlebotomy every 6 months. Arterial blood gas analysis showed a pH of 7.42, a PaO₂ of 51 mmHg, and a PaCO₂ of 30 mmHg, consistent with chronic hypoxaemia and compensated respiratory alkalosis. Chest radiography showed dextrocardia with situs inversus, mild cardiomegaly, and slightly reduced pulmonary vascular markings, with no evidence of pulmonary venous congestion or pleural effusion. Electrocardiography showed sinus rhythm at 78 beats per min, an electrical axis of +15°, a PR interval of 160 ms, and a P-wave duration of 120 ms with an amplitude of 2.5 mm and morphology consistent with atrial enlargement. The P wave was negative in lead I, and features of dextrocardia were present. QRS duration was 80 ms, with appropriate R-wave progression in the precordial leads and electrocardiographic evidence of ventricular hypertrophy. Non-specific repolarisation abnormalities were observed, with no signs of acute ischaemia (**Figure 2**).

Transthoracic echocardiography identified mirror-image atrial arrangement consistent with situs inversus. The atrioventricular connections showed a double inlet to a dominant single ventricle of indeterminate morphology, with both atrioventricular valves opening into the same ventricular chamber. A contralateral rudimentary ventricular chamber was also identified. The ventriculoarterial connections were discordant, consistent with TGA, with the aorta arising from the dominant ventricle. The aortic outflow tract was mildly dilated, with preserved systolic flow. Significant pulmonary



Figure 1. Clinical photograph of the patient's hand during outpatient assessment, showing digital clubbing characterised by widening of the distal phalanges and increased nail curvature. These findings are consistent with chronic hypoxaemia secondary to complex cyanotic congenital heart disease.

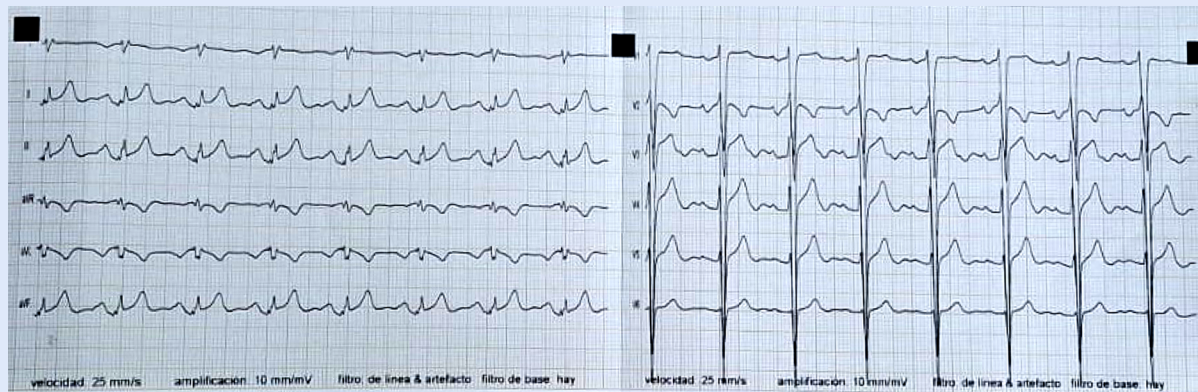


Figure 2. Twelve-lead electrocardiogram.

valve stenosis was documented, with turbulent flow, a peak velocity of 3.9 m/s, and a peak gradient of 60 mmHg, consistent with moderate-to-severe pulmonary stenosis. A large ventricular septal defect and a small atrial septal defect were also identified, both with bidirectional flow. Crossed atrioventricular axes were consistent with criss-cross heart. Global systolic function of the single ventricle was mildly reduced, with an estimated ejection fraction of 50% (**Figure 3**). Advanced investigations, including cardiac magnetic resonance imaging (MRI), cardiovascular computed tomography (CT), and cardiac catheterisation, were not performed during the assessment because of limited access and cost constraints.

The final diagnosis was established by integrating the clinical, electrocardiographic, and echocardiographic findings and was subsequently discussed by specialists during a multidisciplinary assessment. They concluded that the patient had a double-inlet single ventricle associated with TGA, situs inversus, and criss-cross heart. He continued to receive aspirin 100 mg three times per week as preventive therapy in the setting of complex cyanotic CHD, chronic hypoxaemia, and secondary erythrocytosis. He also underwent therapeutic phlebotomy every 6 months to manage secondary erythrocytosis, particularly when the haematocrit exceeded 65% or symptoms suggestive of hyperviscosity syndrome developed.

During 18 months of clinical follow-up, the patient remained stable with conservative management and in NYHA functional class II. He did not require home oxygen therapy and continued to avoid strenuous physical exercise. No hospitalisations for cardiovascular decompensation, syncopal episodes, thromboembolic events, or clinically significant arrhythmias were recorded. Resting oxygen saturation remained stable, and adherence to medical treatment and regular cardiology follow-up was good.

Discussion

SV physiology encompasses a heterogeneous group of complex CHDs characterised by a single functional ventricular

chamber supporting both the systemic and pulmonary circulations. Although CHDs affect approximately 0.8% of liveborn infants, lesions with SV physiology account for a much smaller proportion, with an estimated incidence of less than 1 per 10,000 live births. Population-based studies have reported a prevalence of approximately 16 cases per 100,000 children and only 2 per 100,000 adults, reflecting the high mortality associated with these malformations and the need for early intervention in most patients ^(4,7,8).

The anatomical combination observed in our patient is exceptionally rare. The coexistence of a double-inlet single ventricle, TGA, situs inversus, and criss-cross heart is rarely documented. Criss-cross heart is itself an uncommon malformation, characterised by crossing of the atrioventricular axes resulting from abnormal cardiac rotation during embryonic development. When this anomaly coexists with laterality abnormalities, such as situs inversus, and complex ventriculoarterial defects, anatomical delineation generally requires comprehensive multimodality assessment ⁽⁹⁻¹³⁾.

From a pathophysiological perspective, survival in patients with SV physiology depends primarily on the balance between the systemic and pulmonary circulations. Obligatory mixing of oxygenated and deoxygenated blood results in varying degrees of chronic hypoxaemia, while volume overload of the dominant ventricle promotes progressive heart failure. Consequently, most patients require palliative interventions during childhood to optimise haemodynamics and improve long-term survival ^(4,7,14).

The current standard of care is a staged surgical strategy culminating in Fontan circulation. This usually begins with early palliative procedures, followed by a bidirectional Glenn superior cavopulmonary shunt and subsequent Fontan completion. These interventions aim to progressively separate the systemic and pulmonary circulations, reduce SV volume overload, and improve systemic oxygenation. Contemporary outcomes have improved substantially, with perioperative mortality below 2% in specialist centres ^(6,14-17).

The particular interest of our case, however, lies in the absence of surgical intervention. Survival into the third decade of life in patients with an unoperated single ventricle is uncommon

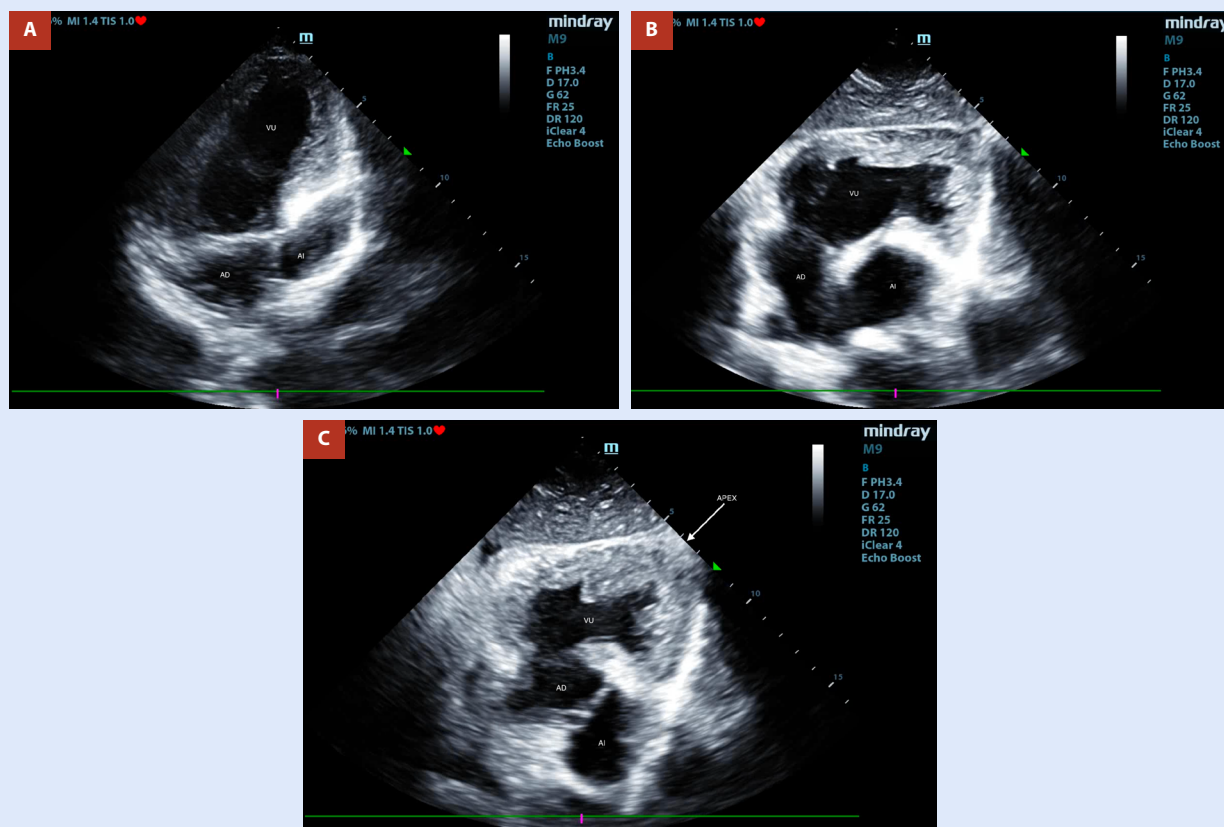


Figure 3. Two-dimensional transthoracic echocardiography. **A)** Apical four-chamber view showing a dominant SV with an enlarged cavity and a hypertrophied wall. **B)** Subcostal view showing concentric hypertrophy of the SV with rudimentary trabeculations. **C)** Subcostal view showing both atria connected to the SV through functional atrioventricular valves, consistent with a double-inlet SV. Echocardiography also demonstrated situs inversus, transposition of the great arteries, and a criss-cross cardiac configuration. Global single-ventricle systolic function was mildly reduced, with an estimated ejection fraction of 50%. Moderate-to-severe pulmonary valve stenosis was documented, with a peak velocity of 3.9 m/s and an estimated peak gradient of 60 mmHg.

SV: single ventricle.

and has been described mainly in small series and isolated case reports. Zheng *et al.* studied patients with SV physiology who had not undergone Fontan palliation and found that some achieved prolonged survival owing to favourable anatomical and haemodynamic characteristics ⁽⁵⁾. Similarly, Gordon *et al.* showed that patients with a single ventricle who have not undergone Fontan palliation constitute a heterogeneous population, in whom survival is closely related to the residual haemodynamic balance and underlying physiology ⁽¹⁾.

Several authors have suggested that moderate obstruction to pulmonary blood flow can have a protective effect by limiting pulmonary overcirculation and preventing the early development of irreversible pulmonary vascular disease. This mechanism appears particularly relevant to our patient, who had moderate-to-severe pulmonary stenosis. This obstruction probably helped maintain a favourable balance between systemic and pulmonary blood flow, preventing both pulmonary overcirculation and early deterioration of ventricular function ^(1,5,13).

The relatively preserved SV systolic function, absence of significant atrioventricular valve regurgitation, and absence of arrhythmias, syncope, or thromboembolic events probably also contributed to the observed clinical stability. These findings are consistent with factors associated with a better prognosis in cohorts of adults with unoperated SV physiology ^(1,5).

Despite the favourable course observed to date, these patients remain at risk of potentially serious late complications, including progressive heart failure, secondary erythrocytosis, hyperviscosity syndrome, arrhythmias, thromboembolic events, and deterioration of ventricular function. Current guidelines therefore recommend lifelong specialist follow-up by multidisciplinary teams with expertise in adult CHD ^(1,5,14).

In conclusion, this case documents the natural history of an exceptionally rare anatomical combination of a double-inlet single ventricle, TGA, situs inversus, and criss-cross heart in an adult who had not undergone surgical palliation (**Figure 4**). Survival to this age probably reflects a favourable haemodynamic balance maintained over many years,

providing valuable insight into the natural history of complex CHD in patients who rarely reach adulthood without surgical intervention.

Ethical aspects

Before preparation of this case report, informed consent was obtained from the patient, ensuring that he understood the information provided and gave consent voluntarily. Confidentiality and anonymity were maintained by omitting

personally identifiable information in accordance with ethical principles.

Author contributions

BVL and JBA: Conceptualisation, study design, investigation, data analysis and interpretation, writing- original draft, and critical review and editing of the manuscript. **SGG, EVL, and AAP:** Investigation, data collection, critical review of the manuscript, and approval of the final version.

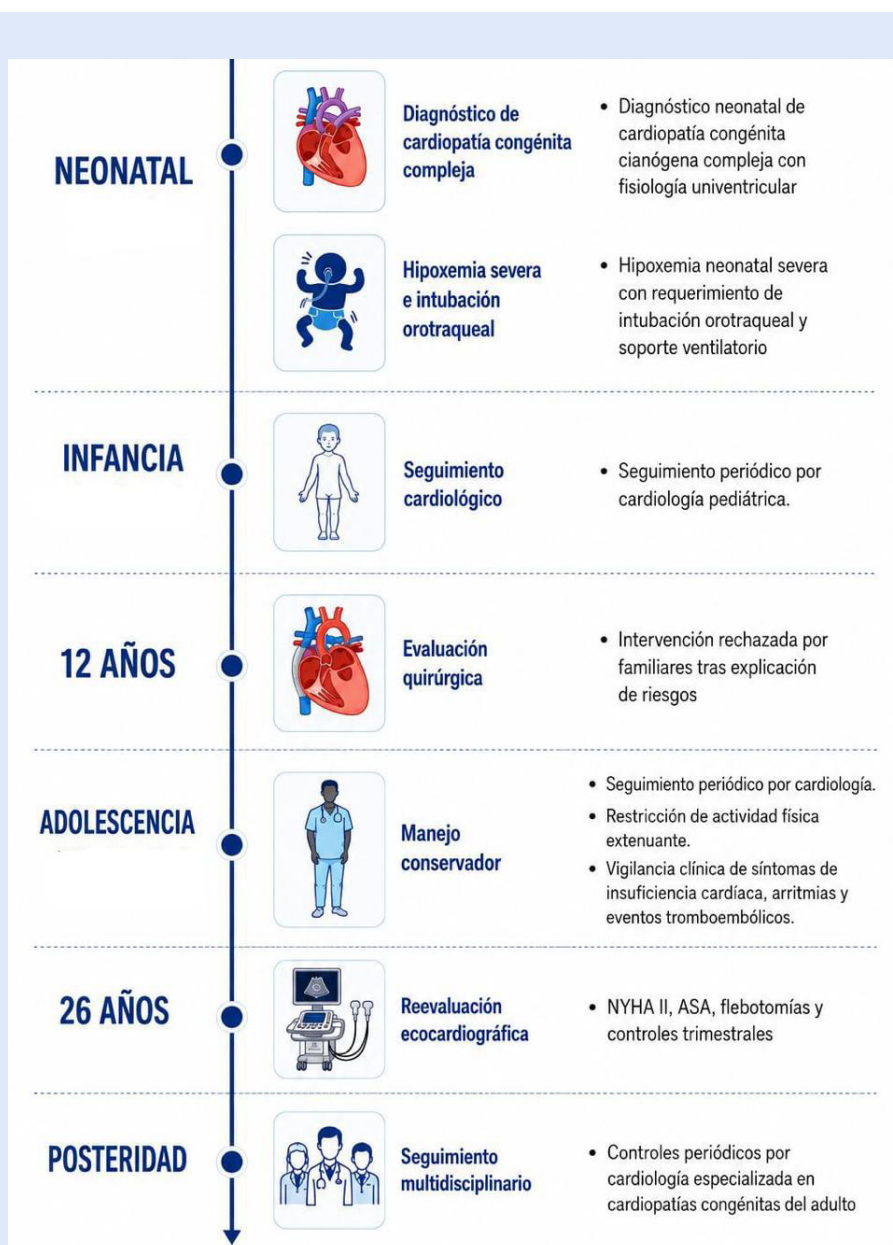


Figure 4. Timeline of the main clinical, diagnostic, and therapeutic milestones during the patient's course.

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