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Case Report

Cardiocentro ERNESTO CHE GUEVARA SANTA CLARA CUBA

# *Echocardiographic diagnosis of persistent truncus arteriosus in an adult patient*

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ARTICLE INFORMATION

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**Competing interests**
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**ABSTRACT**
Persistent truncus arteriosus is a rare form of congenital heart disease. Without surgical repair, only 25% of patients survive the first year of life, and most die during the first three months. The existence of patients who survive to adulthood without surgical correction has been reported, but it is unusual. The case of a 24-year-old male patient, with a previous diagnosis of primary pulmonary hypertension, is reported here. Transthoracic echocardiography showed a persistent truncus arteriosus, type I, in its natural course, with systolic-diastolic dysfunction of both ventricles and moderate insufficiency of the tricuspid and truncal valves. Surgical treatment was contraindicated; therefore, medical treatment started. The patient showed a good general condition at two years of clinical and echocardiographic follow-up. The diagnosis of complex congenital heart disease is not unique to pediatric patients. Transthoracic echocardiography is useful for making an accurate diagnosis without using other diagnostic techniques.
***Key words:*** Congenital heart disease in adults, Persistent truncus arteriosus, Echocardiography

On-Line Versions:
Spanish - English

## *Diagnóstico ecocardiográfico de tronco arterioso común en un paciente adulto*

**RESUMEN**
El tronco arterioso común es una cardiopatía congénita troncoconal poco frecuente. Sólo 25 % de los pacientes sin reparación quirúrgica sobreviven el primer año de vida, la mayor parte fallece durante los primeros tres meses. Se ha registrado la existencia de pacientes sin corrección quirúrgica que sobreviven hasta la edad adulta, pero no es lo usual. En este artículo se presenta un paciente de 24 años de edad con diagnóstico previo de hipertensión pulmonar primaria. En ecocardiograma transtorácico se diagnostica un tronco arterioso común tipo I, en evolución natural, con disfunción sistodiastólica de ambos ventrículos e insuficiencia moderada de las válvulas tricúspide y

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troncal. Existe contraindicación para tratamiento quirúrgico. Se inicia tratamiento médico. A dos años de seguimiento clínico y ecocardiográfico el paciente muestra buen estado general. El diagnóstico de cardiopatías congénitas complejas no es exclusivo de edades pediátricas. La ecocardiografía transtorácica es útil para realizar un diagnóstico certero sin necesidad de emplear otros medios diagnósticos.
***Palabras clave:*** Cardiopatía congénita del adulto, Tronco arterioso común, Ecocardiografía

## INTRODUCTION

Persistent truncus arteriosus is an unusual form of congenital heart disease, which accounts approximately for 1 to 5% of all congenital heart diseases[1]. It is characterized by a single arterial trunk that emerges from the heart and gives rise to systemic, pulmonary and coronary circulation[2]. Collett and Edwards classification[3] has been the most commonly used one, and describes four types. In type I, a short main pulmonary artery originates from the common trunk and forms both pulmonary arteries. Surgical correction is undertaken during childhood, and exceptionally in adulthood. Without surgical repair, only 25% of patients survive the first year of life, and most die during the first three months. The existence of patients who survive to adulthood without a correction of the truncus arteriosus is known, but it is not common. Transthoracic echocardiography is the noninvasive method of choice for establishing a diagnosis[4,5].

## CASE REPORT

A 24-year-old male patient came to hospital with a diagnosis of probable primary pulmonary hypertension.

The patient complained of dyspnea on moderate exertion. Hyperdynamic precordium, clubbing and cyanosis were found during examination. The respiratory rate was 26 breaths per minute. On auscultation, a continuous murmur, grade 3-4, was detected in the cardiac base, accompanied by a thrill and a heart rate

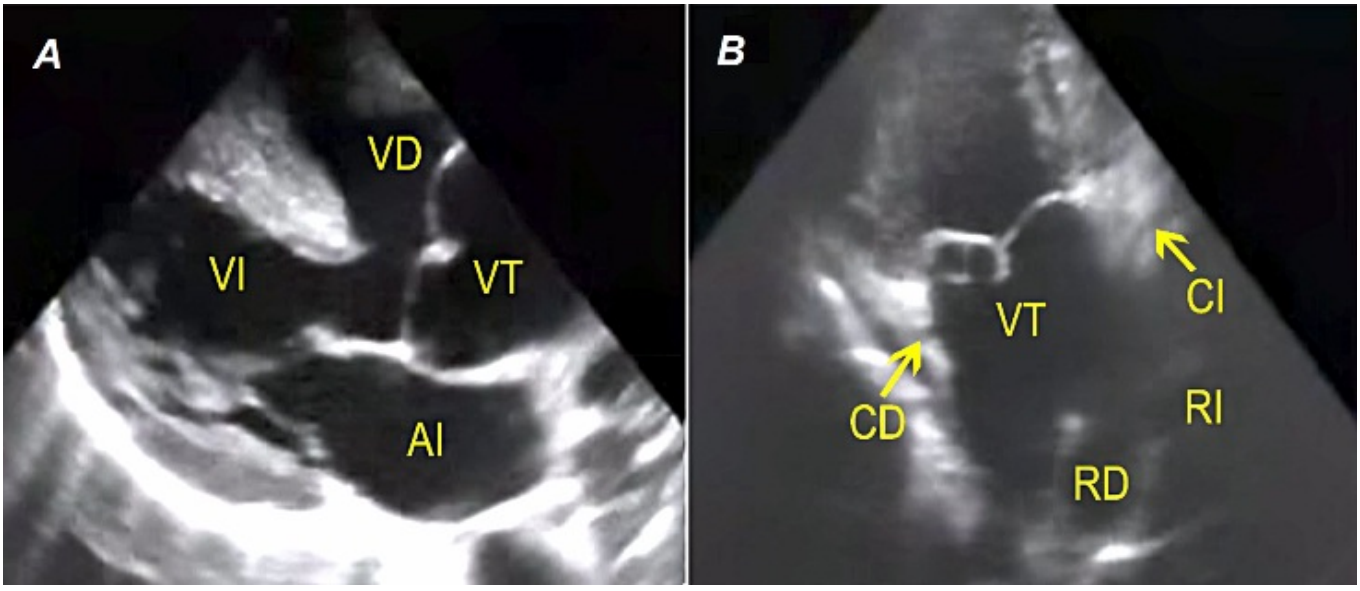


**Figure 1. A.** Parasternal long axis view, where the trucal vessel overrides the large conoventricular ventricular septal defect. **B.** (Persistent truncus arteriosus, type I) Pulmonary arteries originating from the left posterolateral wall of the truncal vessel by a short pulmonary trunk, right and left coronary arteries emerging from right and left coronary sinuses. Acronyms in Spanish - AI: left atrium, CD: right coronary artery, CI: left coronary artery, RD: right pulmonary artery, RI: left pulmonary artery, VD: right ventricle, VI: left ventricle, VT: Truncal vessel.

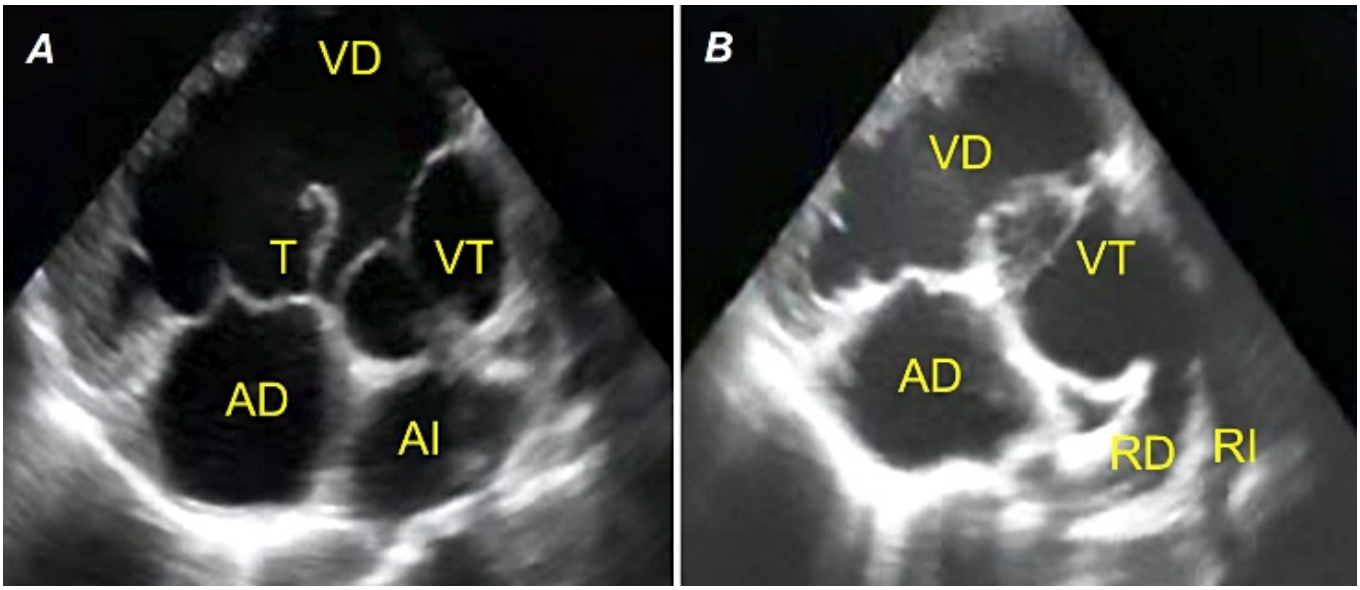


**Figure 2. A.** High parasternal short axis view, where the truncal vessel is seen, as well as dilated right ventricle and accessory tricuspid tissue. **B**. Persistent truncus arteriosus, type I. Acronyms in Spanish - AD: right atrium, AI: left atrium, RD: right pulmonary artery, RI: left pulmonary artery, T: accessory tricuspid tissue, VD: right ventricle, VT: truncal vessel.

of 78 beats per minute. Arterial oxygen saturation was 68%. Peripheral arterial pulses were bounding. The electrocardiogram showed biventricular hypertrophy and the telecardiogram indicated a severe cardiomegaly.

Situs solitus was shown in the two-dimensional transthoracic echocardiography, with atrioventricular concordance, moderate dilatation of both ventricles, and the truncal vessel overriding the crest of the interventricular septum, where a large conoventricular ventricular septal defect is located (**Figure 1A**). A type I persistent truncus arteriosus was identified. In it, coronary circulation and both pulmonary arteries emerge from a short pulmonary trunk (**Figures 1B and 2B**). Accessory tricuspid tissue was found, as well as moderate insufficiency of the tricuspid and truncal valves (**Figure 2A**). There was a severe biventricular systolic dysfunction. Multiple systemic-pulmonary collaterals were detected, and elevated pulmonary pressures; therefore surgical reconstruction was contraindicated. Medical treatment was started with sildenafil, carvedilol and spironolactone in normal doses.

After two years of clinical follow-up, the patient showed a good general condition, and subsequent echocardiograms have shown a recovery of biventricular systolic function, with regression of initial ventricular diameters.

## COMMENT

This case demonstrates the importance of transthoracic echocardiography for adult patients with complex congenital heart disease. To our knowledge, without surgical correction, the prolonged survival of patients with persistent truncus arteriosus has not been described. Clinical symptoms are not specific at this stage of life, and the clinical findings of pulmonary hypertension secondary to congenital heart disease often predominate. Electrocardiographic and radiological findings are also nonspecific; therefore, the two-dimensional transthoracic echocardiography is crucial for establishing a diagnosis. The views of the parasternal long axis and the high parasternal short axis[4,5] were used (**Figures 1 and 2**).

It is essential to have a high level of suspicion when assessing an adult patient with cyanosis, because age is not a factor that excludes the diagnosis of complex congenital heart disease. It is considered that transthoracic echocardiography is useful for making an accurate diagnosis without using other diagnostic techniques.

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