CASE REPORT

Late correction of transposition of great arteries in a 14-year-old child

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Transposition of the great arteries (TGA) has a high mortality if not treated early. There are presentations where wide interventricular or atrial septal defect allow greater mixing, manifesting a lighter clinic picture. In these cases, the diagnosis is usually made late. We present the case of a 14-year-old female with transposition of the great arteries and pulmonary stenosis who underwent a successful arterial switch with implantation of a biological prosthesis in the neoaorta.

***Key words:** Arterial Switch; Aortic valve replacement; Congenital Heart Disease; Transposition of the Great Arteries.*

La transposición de las grandes arterias tiene una mortalidad alta si no se trata de manera temprana. Existen presentaciones donde la comunicación interventricular o interauricular amplia permiten una mayor mezcla, manifestando un comportamiento clínico mas leve. En estos casos el diagnóstico suele hacerse de manera tardía. Se presenta el caso de una femenino de 14 años con ransposición de las grandes arterias con estenosis pulmonar a quien se le realizó de manera exitosa un switch arterial con implante de prótesis biológica en neoaorta.

***Palabras clave**: Switch arterial; Reemplazo valvular aórtico; Cardioptía congénita; Transposición de las Grandes Arterias.*

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Transposition of the great arteries (TGA) is the most frequent cyanotic congenital heart disease in the neonatal stage. It is characterized by an anterior aorta to the right of the pulmonary artery. Without surgical treatment, 89.3% do not survive the first year [1]. It is uncommon to see a patient older with this pathology due to hemodynamic complications. However, certain anatomical characteristics such as pulmonary stenosis and large ventricular septal defect (VSD) can contribute to a late presentation of TGA [2]. A case is presented in an adolescent.

CLINICAL CASE

A 14-year-old female with a late diagnosis of TGA with pulmonary valve stenosis and interventricular septal defect was presented. A murmur was detected at 8 months of life, without follow-up. Subsequently, the patient presented clubbing of fingers and cyanosis treated by hemodilution due to polycythemia on 3 occasions. As a part of the diagnostic approach in our unit, a transthoracic echocardiogram was performed (**Fig. 1**) (**Fig. 2**) (**Fig. 3**) (**Fig. 4**). This study demonstrated situs solitus, atrioventricular concordance, and ventriculoarterial discordance with pulmonary stenosis. Cardiac catheterization reported TGA with pulmonary valve stenosis, interventricular septal defect, pulmonary valve stenosis, small atrial septal defect, and direct aorto -pulmonary collaterals. The decision-making process was in favor of undergoing surgery.

Arterial switch (Jatene), Inspiris Resilience Aortic Valve implantation in neoaorta and VSD closure with bovine pericardium patch were performed at the same time. The course in ICU was uneventful, with a length of stay of 6 days. Vasoactive support was administered along 2 days, and the extubation took place at 48 hours after operation. Clinically with improvement of functional class and saturation 90-94% was observed, leading to a satisfactory hospital discharge.

COMMENT

TGA occurs in 1 in 3,000 live births. In the simple form of the disease, the integrity of the interventricular septum exists and there is no obstruction of the left ventricular outflow tract, usually presenting more marked cyanosis [1] that requires prompt access to a surgical center [3].

In TGA without surgical treatment, 28.7% die in the first week of life, 51.6% in the first month, and 89.3% do not survive the first year [1]. Surgical treatment can be performed depending on the severity by palliation, preparation of the

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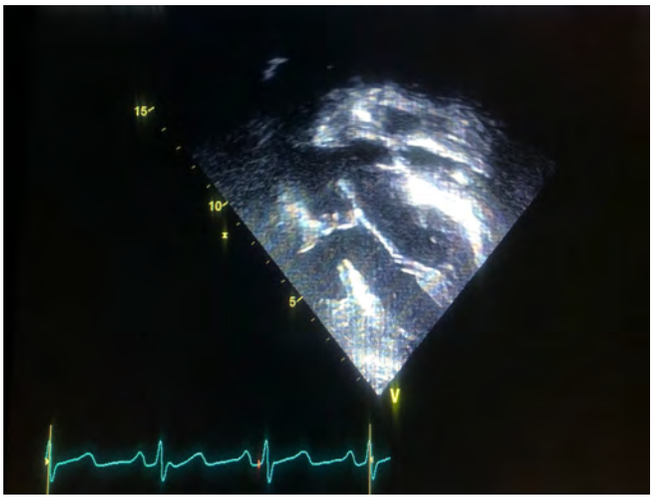

Figure 1. Echocardiogram image with five-chamber apical axis. Pulmonary artery emerging from the left ventricle with a defect at the level of the interventricular septum is observed.

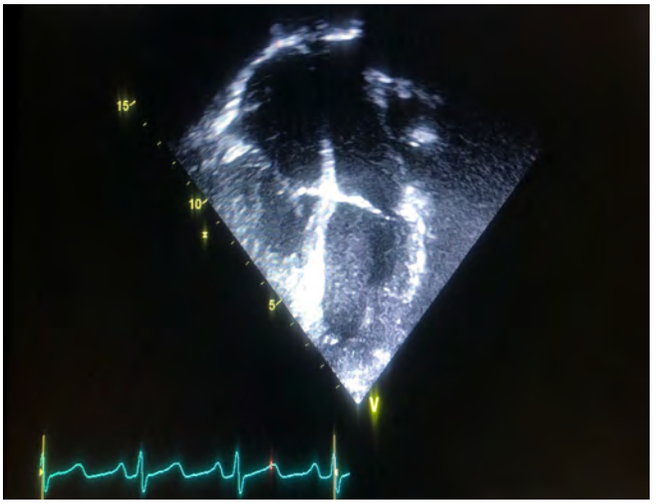

Figure 3. Four-chamber echocardiogram image showing concordant atrioventricular connection, dilatation of the left cavities, and concentric hypertrophy of the right ventricle.

left ventricle, Senning, Mustard or by arterial switch. This last procedure replaced palliative surgeries in patients with TGA since it was performed for the first time in Sao Paulo in 1975 by Abid Jatene [2].

In the most complex form of the disease, it is associated with VSD, pulmonary stenosis and coarctation of the aorta. Depending on the combination of these abnormalities, the hemodynamic presentation may vary. For example, small ventricular and atrial septal defects are associated with more severe symptoms and vice versa. As for pulmonary stenosis, it can worsen the condition and the prognosis [1].

Late arterial switch is considered in a child older than 3 or 4 weeks of age [3]. However, there are certain cases where the patient survives due to a VSD or large atrial septal defect. The clinical picture is softer and cyanosis may not be so evident, which leads to a late diagnosis. Usually, these children have manifestations of pulmonary hyperflow [4]. In the case of our patient, the absence of left ventricular obstruction, coupled with pulmonary stenosis, could have contributed to the absence of more severe pulmonary hypertension, which influenced her survival [1].

In any case, diagnostic suspicion is important for early detection. The gold standard is echocardiography in expe-

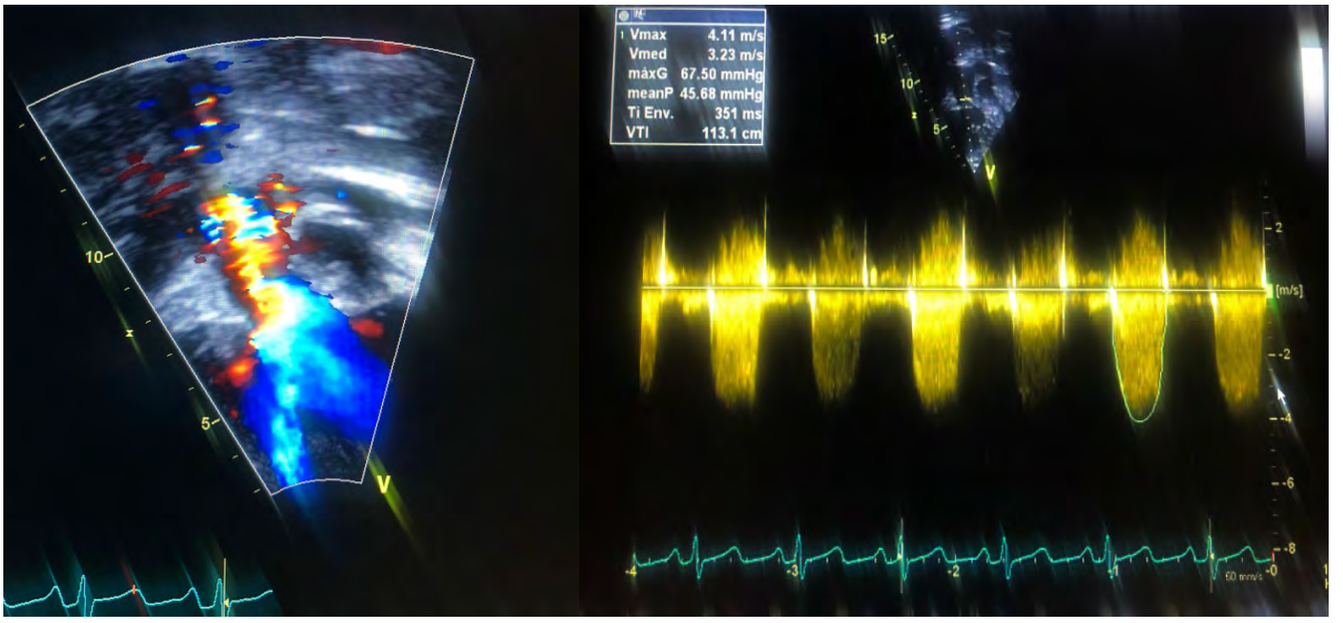

Figure 2. A and B. Image with apical 5-chamber view with color Doppler showing anterograde flow with a peak gradient of 67mmHg and a mean gradient of 45mmHg.

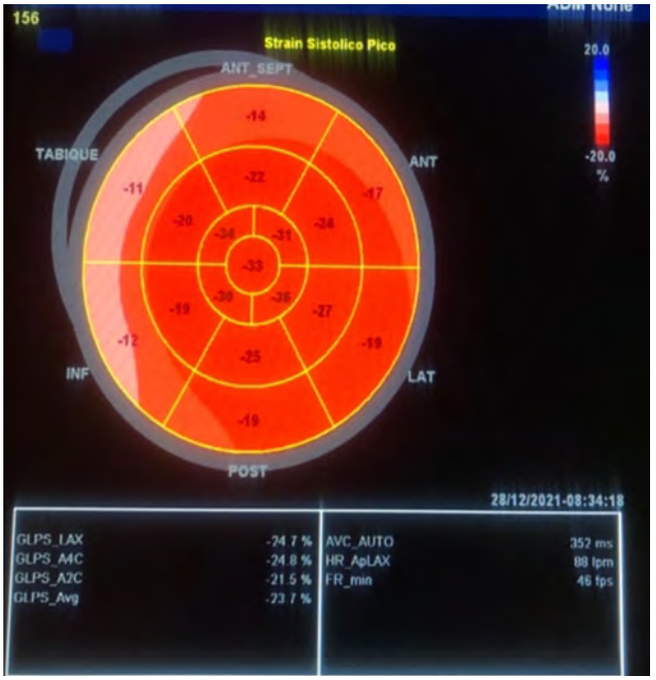

Figure 4. Figure 4. Ventricular systolic function by transthoracic echocardiogram in a 14-year-old adolescent with TGA: LV global strain -23.7% with a greater area of deformation at the level of the ventricular septum.

rienced hands, which should show atrial situs, atrioventricular concordance and ventriculoarterial discordance, and of course associated anomalies such as VSD or pulmonary stenosis. Cardiac catheterization is preferable in older patients where pulmonary vascular disease is suspected [4].

An arterial switch was performed on our patient that also required the implantation of a biological valve prosthesis in the neoaorta due to the presence of pulmonary stenosis, with an excellent postoperative evolution. However, it should be considered that patients corrected with VSD have a higher rate of reintervention [5], due to associated injuries. Also, as in this case due to the presence of a biological valve.

The prognosis of patients with arterial switch at this age has not been reported due to the shortage of cases. However, a recent study reported that patients over 6 months of age at the time of arterial switch had quality of life scores indistinguishable from their peers counterparts [5].

Finally, it is important to mention that if the correction has not been performed, patients with large VSD may present hepatomegaly and symptoms of congestive heart failure.

In conclusion, The absence of left ventricular obstruction and a large VSD allows adequate mixing between the two circulations that allows detection at this age. Anatomical correction can be carried out in some cases.

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