Gac Méd Caracas 2021;129(1):167-171
DOI: 10.47307/GMC.2021.129.1.17

Five cardiopathies, one heart

Cinco cardiopatías, un corazón

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SUMMARY

This clinical case reports a patient diagnosed with a partial 1- Cor Triatriatum Sinister associated with 2- severe calcified mitral stenosis and accessory subvalvular mitral tissue, 3- membranous subvalvular aortic stenosis, 4- severe aortic valve regurgitation, and 5- patent conduct arteriosus (PCA), with mild depressed ejection fraction and moderate pulmonary hypertension who underwent surgery for successful membrane resection and corresponding mitro-aortic valve replacement, in addition to closing intracardiac of the patent artery duct. Early and late postoperative evolution was satisfactory. This is the first reported case of this type of congenital heart disease in an adult in Venezuela.

Key words: *Cor triatriatum, subvalvular aortic stenosis, aortic valve regurgitation, patent conduct arteriosus.*

RESUMEN

El presente caso clínico trata sobre un paciente con diagnóstico de 1- Cor Triatriatum Sinister parcial asociado a 2. Estenosis mitral severa calcificada y tejido subvalvular mitral accesorio, 3. Estenosis subvalvular aórtica membranosa, 4. Insuficiencia valvular aórtica severa y 5. Persistencia del conducto arterioso (PCA), con fracción de eyección deprimida leve e hipertensión pulmonar moderada. Se le realizó de manera exitosa resección de las membranas, además del cierre intracardíaco del conducto arterioso y reemplazo valvular mitro-aórtico. La evolución posoperatoria temprana y tardía fue satisfactoria. Se trata del primer caso reportado de este tipo de cardiopatías congénitas en el adulto en Venezuela.

Palabras clave: *Cor triatriatum, estenosis aórtica subvalvular, insuficiencia valvular aórtica, conducto arterioso permeable.*

INTRODUCTION

First reported in 1868, cor triatriatum, that is, a heart with 3 atria (triatrial heart), is a congenital anomaly in which the left atrium (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is divided into 2 compartments by a fold of tissue, a membrane, or a fibromuscular band (1).

The embryologic basis of this condition is unclear and controversial. Different theories had attributed this anomaly to mal

DOI: https://doi.org/10.47307/GMC.2021.129.1.17

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Recibido: 20 de noviembre 2020
Aceptado: 4 de enero 2021

septation, mal incorporation, or entrapment during cardiac development (2-4). Lewis first reported operative treatment of cor triatriatum in 1956 (5). Resection of cor triatriatum membrane using cardiopulmonary bypass is the definitive treatment. The majority of patients have associated congenital cardiovascular anomalies (6-8). The surgical outcome depends upon the complexity of associated cardiac anomalies (9,10).

In high volume echocardiographic laboratories, the incidence of cor triatriatum is less than 1 in 10 000. However, this is expected to rise with the increasing use of cardiac diagnostic studies counting for 0.1 % of all congenital cardiopathies (1).

In symptomatic infants, cortriatriatum sinistrum (CTS) is often associated with other major congenital cardiovascular defects. In the adult, CTS can be as follows (1):

Asymptomatic (found incidentally on cardiac imaging).

An isolated finding with a large nonrestrictive communication between the superior and inferior left atrial chambers.

Associated with minor congenital defects such as patent foramen ovale, atrial septal defect, pulmonary anomalous drainage, or persistent left superior vena cava.

Diagnosis has become increasingly accurate thanks to advances in imaging studies, and today two-dimensional echocardiography and color Doppler have become the non-invasive diagnostic modality of choice since the 1980s (11).

There are few centers with reported series of patients involved with this pathology, with periods spanning up to 50 years and including pediatric and adult patients as the series reported by The Mayo Clinic (12); In the adult, the most frequently reported are isolated cases, mainly associated with the closure of interatrial septum defect, correction of abnormal venous drainage, valvular replacements for degenerative disease, among others (13-16).

In the literature reviewed, in Venezuela there were found no cases published with this pathology, being this perhaps the first case in adults published to date.

CASE REPORT

This is a 30 years old male patient, from the interior of the country, with a cardiopathy diagnosis since childhood, with eventual medical controls in JM de Los Ríos Hospital, and an effort dyspnea progression for 3 months before he consults to Military Hospital of Caracas from which he is referred to University Hospital of Caracas, being admitted in May of 2015 for a joint evaluation between Cardiology and Cardiovascular Services.

Thoracic X-ray showed only unspecific findings such as augmented cardiothoracic index, probably due to the left atrium and ventricle enlargement, a prominent pulmonary artery, and hyper flux in pulmonary fields.

A transthoracic echocardiogram is performed evidencing an acyanotic congenital cardiopathy evidencing a Patent Conduct Arteriosus (PCA) of small size, with an estimated diameter of 0,5 cm and left to right shunt. Aortic valve with high gradients probably due to jet acceleration related to a subvalvular membrane and moderate regurgitation. Left ventricle severely dilated with severe eccentric hypertrophy and mild diffuse hypokinesis, mildly depressed systolic function, the ejection fraction of (EF) 45%. Severe mitral valve stenosis, probable parachute mitral valve, maximum velocity (maxV) 2,17m/s, mean gradient (mGrad) 10,1 mmGg, mitral valve area by continuity equation 0.95 cm^2, and mild insufficiency. Moderate pulmonary valve insufficiency. Mild tricuspid valve insufficiency with moderate pulmonary artery hypertension (PAH) 45-50 mmHg. Severely dilated right ventricle with normal systolic function (AF 35 %). Pulmonary artery and its branches dilated.

Because of these findings, aortic subvalvular anomalies and mitral valve characteristics were preceded by transesophageal echocardiography showing sclera calcificated aortic valve with high gradients (mGrad=36 mmHg), probably related with a subvalvular membrane sided towards the interventricular septum besides the right coronary leaflet (Figure 1) also with a severe insufficiency due to coaptation failure of the leaflets with Vena Contracta (VC) of 7 mm. A calcified and stenotic mitral valve. A supravalvular membrane and a medium auricular membrane were observed in the left atrium (Figure 2).

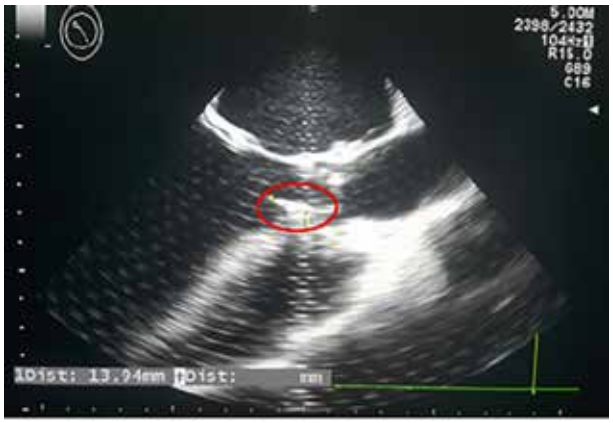

Figure 1. Subvalvular aortic tissue membranous type detail sized 13,94 mm (Red circle).

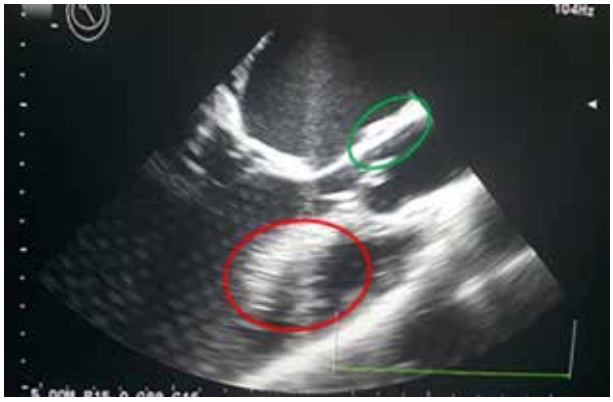

Figure 2. A 3-chambers transesophageal. Echocardiography view where it shows a thickened with low implantation mitral valve with subvalvular accessory tissue (red circle) and a partial view of a calcified supravalvular mitral ring (green circle).

A right cardiac catheterization is decided for pressure registry concluding PCA and severe PAH.

In a joint meeting between Cardiovascular Surgery Service, Cardiology Service, and Congenital & Pediatric Cardiopathies Section, it was questioned whether the patient still was in the surgical window, deciding in a Cardiovascular Service reunion to offer a more aggressive surgical resolution due to the patients' age and because of the impossibility of a heart transplant in the country.

Surgery was performed on August 7th of 2015, by a mean sternotomy approach, with aortic and bicaval cannulation, the entry in extracorporeal circulation, and aortic cross-clamping. Through right atriotomy introducing retroplejia cannula under direct view inside the coronary sinus and then transeptal approach the left atrium and mitral valve was accessed. PCA was identified and dissected, then through an arteriotomy in the main pulmonary trunk, and with direct visualization of PCA´s orifice this was closed with a running 5-0 prolene suture. A supravalvular mitral calcified ring (unrestrictive Cor Triatriatum) (Figure 3), anomalous and low implantation with severe stenotic mitral valve were between the intraoperative findings, proceeding to the supravalvular ring (Figure 4) and mitral valve with subvalvular accessory tissue resection, latter pledgetedethibond stitches implantation in the annulus and subsequently a mechanic On-X 27-29 mm mitral valve. On aortotomy became evident lack of coaptation of aortic leaflets and a subvalvular membranous type stenosis, both were resected in the same manner with latter implantation of valve sutures to a mechanic ATS medical 20mm aortic valve in the annulus (Figure 5). A consecutive closure of transeptal, right atrium, and aortotomy approach was performed with aerial emboli preventive maneuvers. Extracorporeal circulation time was 3 hours and 36 minutes and aortic cross-clamping time of 2 hours and 32 minutes. The postoperative diagnosis was as follows: Partial Cor Triatriatum Sinister, Severe calcified mitral stenosis, subvalvular accessory mitral tissue, Subvalvular Aortic Stenosis, and severe aortic valve insufficiency, and finally Patent Conduct Arteriosus. With satisfactory postoperative evolution, extubation accomplished on 12 hours postoperative, discharge from the intensive care unit (ICU) on 48h postoperatively, and medical discharge from hospital 10 days later, with oral anticoagulation and outpatient medical management to present day.

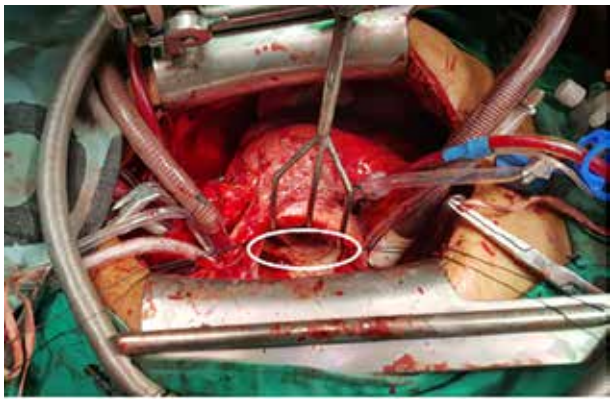

Figure 3. Evidence of thick and calcified mitral supravalvular ring (white circle: supravalvular ring).

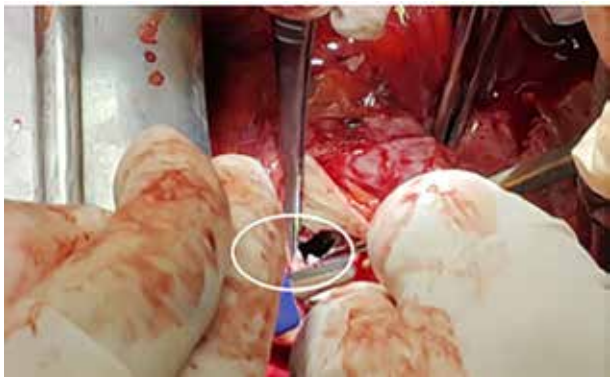

Figure 4. Detail of moment of resection of the mitral supravalvular ring with scalpel.

Figure 5. Mechanical aortic valve implantation.

DISCUSSION

As presented in the reviewed literature, in adults with Cor Triatriatum congenital heart disease associated tend to be minor (1), allowing them to reach adulthood without having undergone surgery in childhood (12-16), however, it should be noted that this same situation coupled with occasional controls may expose the patient to a silent advance in the muscular cardiac dysfunction degree to almost getting inoperable, more so, when the associated heart disease is of valvular etiology, which predisposes them to both degenerative and the possible antecedent of rheumatic disease. In our country the distant domicile, the precariousness, and inadequacy of the health infrastructure, makes it difficult to adequately control and monitor those adult patients with known diagnoses of congenital heart disease, since the few reference centers with the capacity and experience to perform this type of surgery are located in the capital, leaving unattended the country within.

Despite the reserved prognosis and doubts from the Congenital Cardiopathy Section, the successful conclusion of this case demonstrates the importance of interdisciplinary management as well as the necessary surgical experience in a national reference center.

REFERENCES

1. Shirani J. Cor Triatriatum: Background, Pathophysiology, Epidemiology. http://emedicine.medscape.com/article/154168overview#showall.
2. Gharagozloo F, Bulkley B, Hutchins G. A proposed pathogenesis of cortriatriatum: Impingement of the left superior vena cava on the developing left atrium. Am Heart J. 1977;94:618-626.
3. Van Praagh R, Corsini I. Cor triatriatum: Pathologic anatomy and a consideration of morphogenesis based on 13 postmortem cases and a study of normal development of the pulmonary vein and atrial septum in 83 human embryos. Am Heart J. 1969;78:379-405.
4. Fowler J. Membranous band in the left auricle. Trans Pathol Soc Lond. 1881;33:77-94.
5. Lewis F, Varco R, Taufic M, Niazi S. Direct vision repair of triatrial heart and total anomalous pulmonary venous drainage. Surg Gynecol Obstet. 1956;102:713-720.

6. Humpl T, Reineker K, Manlhiot C, Dipchand A, Coles JG, McCrindle BW. Cor triatriatum sinistrum in childhood. A single institution's experience. Can J Cardiol. 2010;26:371-376.
7. Yaroglu Kazanci S, Emani S, McElhinney D. Outcome after repair of cortriatriatum. Am J Cardiol. 2012;109:412-416.
8. Alphonso N, Nørgaard M, Newcomb A, d'Udekem Y, Brizard CP, Cochrane A. Cor triatriatum: Presentation, diagnosis and long-term surgical results. Ann Thorac Surg. 2005;80:1666-1671.
9. Salomone G, Tiraboschi R, Bianchi T, Ferri F, Crippa M, Parenzan L. Cor triatriatum. Clinical presentation and operative results. J Thorac Cardiovasc Surg. 1991;101:1088-1092.
10. Rodefeld M, Brown J, Heimansohn D, King H, Girod DA, Hurwitz RA, et al. Cor triatriatum: Clinical presentation and surgical results in 12 patients. Ann Thorac Surg. 1990;50:562-568.
11. Bolio-Cerdán C, Medina-Andrade A, Romero-Cárdenas C, Ruiz-González G, Luna-Valdez M, González-Peña J. Cor triatriatum sinistrum: estrategia diagnóstica y terapéutica. Bol Med Hosp Infant Mex. 2007;64:29-34.
12. Saxena P, Burkhart H, Schaff H, Daly R, Joyce LD, Dearani JA. Surgical Repair of Cor Triatriatum Sinister: The Mayo Clinic 50-Year Experience. Ann Thorac Surg. 2014;97:1659-1663.
13. Wong C, Leung M, Cheng M, Lau C, Cheung DL. Myxomatous mitral valve degeneration complicating asymptomatic cor triatriatum. Clin Cardiol. 1989;12:48-50.
14. Barbaglia F, Casanova M, Araujo R, Tazar J. Cor triatriatum sinister. A propósito de un caso. Insuf Card. 2010;5(1):42-47.
15. Eichholz J, Hodroge S, Crook J, Mack J. Cor Triatriatum Sinister in a 43-Year-Old Man with Syncope. Tex Heart Inst J. 2013;40(5):602-605.
16. Chen Q, Guhathakurta S, Vadalapali G, Nalladaru Z, Easthope RN, Sharma AK. Cor Triatriatum in Adults. Three New Cases and a Brief Review. Tex Heart Inst J. 1999;26:206-210.