CASE REPORT

Surgical outcome of a rare case: cor triatriatum dexter

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Cor triatriatum dexter is defined as the division of the right atrium into two compartments due to an embryological defect, accounting for 0.01% of congenital heart diseases. The clinical presentation is variable and the preferred diagnostic methods are ultrasound and MRI. Treatment in symptomatic cases is done by surgical or percutaneous approach. We present the case of a 9-year-old male patient diagnosed with cor triatriatum dexter and atrial septal defect.

Key words: **Atrial septal defect; Congenital heart disease; Cor triatriatum dexter; Right atrium.**

El cor triatriatum dexter se define como la división de la aurícula derecha en dos compartimientos por un defecto embriológico, representando 0.01% de las cardiopatías congénitas. La presentación clínica es variable y los métodos diagnósticos preferidos son la ecografía y la resonancia magnética. El tratamiento en casos sintomáticos es por abordaje quirúrgico o percutáneo. Presentamos el caso de un paciente masculino de 9 años con diagnóstico de cor triatriatum dexter y comunicación interauricular.

***Palabras clave*: Comunicación interauricular; Cardiopatía congénita; Cor triatriatum dexter; Aurícula derecha.**

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Historically, the division of the left atrium (sinister) creating an accessory chamber was first described in 1868 by Church, and in the case of the right atrial division (dexter), it would be described in 1875 by Rokitansky. However, the term Cor triatriatum was coined by Borst in 1905 [1].

Cor triatriatum is a rare congenital anomaly in which one of the atria is divided into two compartments [2]. This term, also known as "triatrial heart", generally refers to cor triatriatum sinister or "divided left atrium", which is a rare condition with an estimated incidence of 0.1% of all congenital cardiac malformations [3]. Cor triatriatum dexter (CTD) or "divided right atrium (RA)" is an even rarer pathology [2]. Its occurence is unknown, but it is estimated to account for less than 0.01% of congenital heart diseases. Between 1897 and 1972 only 14 cases were reported, of which all but one were identified post mortem. With the development of echocardiography, CTD diagnoses have become more common [4].

However, it is still a pathology that has been little described in academia, hence the interest in reviewing the literature and correlating it with the clinical case described below.

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CLINICAL CASE

A 9-year-old male patient. The patient was admitted to this hospital at two months of age, due to the presence of cyanosis from birth, since then he was kept under surveillance without receiving medical treatment until the time of admission, being in functional class I of the New York Heart Association.

The patient was presented at the medical-surgical session with a diagnosis of CTD with wide atrial septal defect (ASD) and persistent cyanosis. After the physical examination, the patient weighed 26 kg (P10), body surface area 0.99 m2, height 132 cm (P25), heart rate 75 bpm, blood pressure 90/50 mmHg, oxygen saturation 78%. Normal phenotype, cyanosis and hypokratism grade I (**Fig.1A**) (**Fig.1B**), respiratory pattern with mild polypnoea. Hyperdynamic prechord, no murmurs, normal 2nd tone. Hepatic border not present to touch, pulses present and symmetrical in 4 extremities and neurologically intact. In the cabinet studies performed, the following is highlighted: Chest X-ray in situs solitus, levocardia, cardiothoracic index of 0.50, decreased pulmonary flow was observed (**Fig. 2A**). Electrocardiogram with ectopic atrial rhythm, heart rate 75 bpm, P -30 cardiac axis, QRS 0.08 sec, QTc 0.42, presence of left bundle branch block (posterior fascicle) (**Fig. 2B**). Transthoracic echocardiogram in situs solitus, levocardia, normal systemic venous returns, atrioventricular and ventricular arterial concordance,

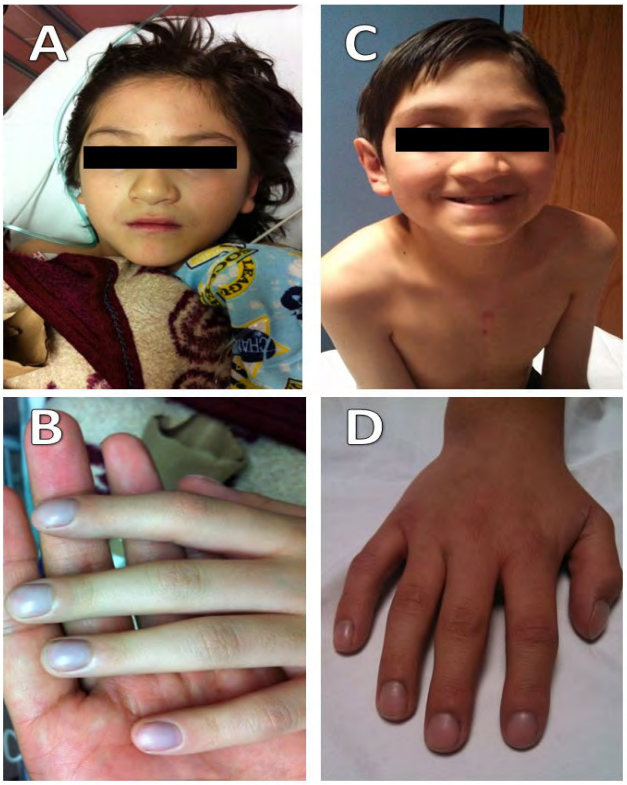

Figure 1. A and B) Patient with cyanosis and preoperative clubbing. C and D) Postoperative patient with remission of clinical manifestations.

19 mm ASD with right-to-left shunt, RA with membrane dividing it into 2 chambers, mild to moderate hypoplasia of the right ventricle (RV), tricuspid annulus hypoplasia, 10 mm right pulmonary artery branch, 9 mm left pulmonary artery branch and 14 mm pulmonary artery trunk (**Fig. 3**). Transesophageal echocardiogram reported CTD with wide ASD.

Surgical findings are described as follows: Resection of the supratricuspid membrane and closure of the ASD (**Fig.4**). An atrial wall bridge (membrane) was noted over the ASD diverting flow from the right pulmonary veins into the tricuspid, ASD at upper septum 15 mm, which was left at 4 mm. A post-surgical echocardiogram was performed, in which no residual interatrial shunt was observed, the function of both ventricles was preserved and the tricuspid valve without insufficiency. The patient had an uncomplicated postoperative period (**Fig. 1C**) (**Fig. 1D**), extubated on the same day of surgery, stayed in the intensive care unit for 3 days and was discharged from the hospital 5 days after surgery.

COMMENT

Cor triatriatum is a congenital anomaly in which one of the atria is divided into two compartments. There are two types: cor triatriatum sinister is the most common form and involves splitting of the left atrium. CTD is even rarer and involves splitting of the RA [2]. CTD is one of the less common congenital cardiac malformations in which there is abnormal RA septation due to a failed regression of the embryonic right sinus venosus valve [5].

Embryologically, the RA develops early in fetal life when the right horn of the venous sinus fuses with the primitive atrium. The venous sinus is a quadrangular cavity preceding the AD. The left horn of the venous sinus becomes the coronary sinus and oblique vein of the left atrium. The right horn carries venous blood from the right anterior cardinal vein (which becomes the superior vena cava), the right vitelline vein (which becomes the inferior vena cava) and the right umbilical vein (which eventually obliterates) to the primitive RA. The sinoatrial orifice, surrounded by the left and right venous valves, is the communication between the right horn of the venous sinus and the primitive RA. The left venous valve eventually fuses with

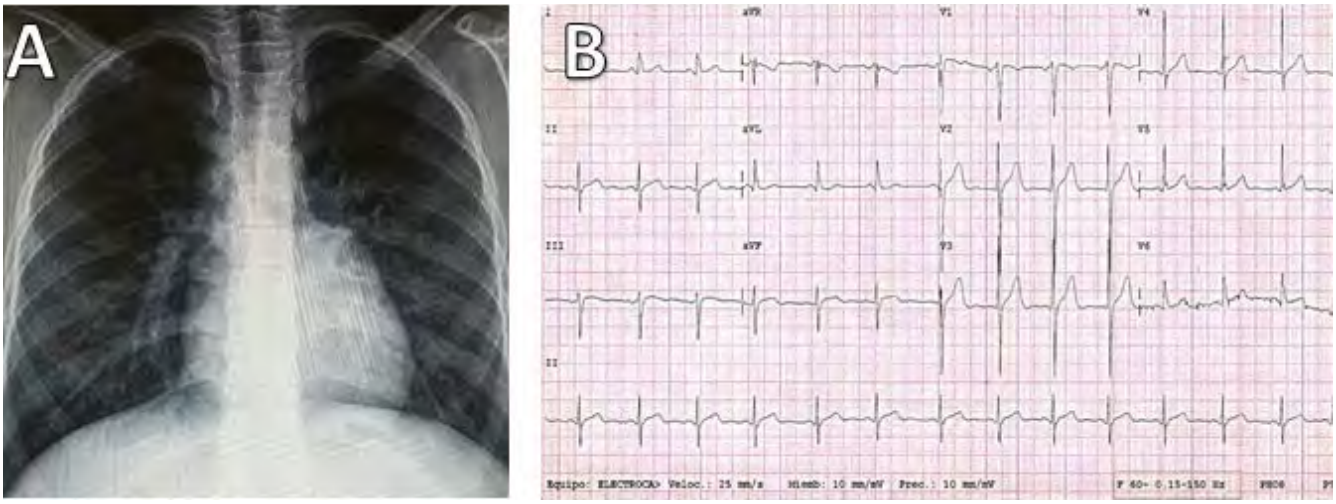

Figure 2. A) Chest X-ray shows decreased pulmonary flow. B) Electrocardiogram with ectopic atrial rhythm and left bundle branch posterior fascicle block.

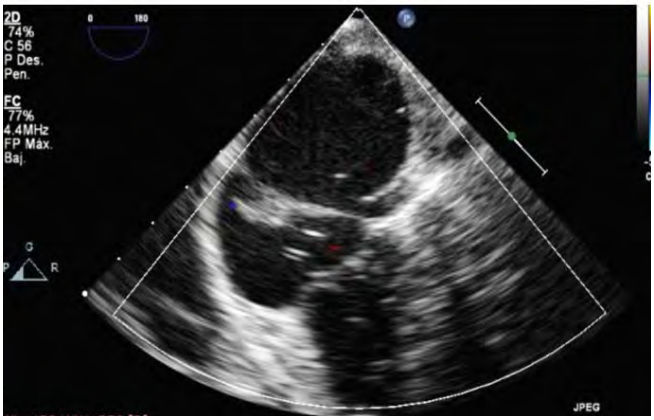

Figure 3. Echocardiogram shows membrane dividing right atrium into two chambers.

the septum secundum, separating the left and right atria. The right venous valve divides the RA into two chambers, allowing oxygenated blood from the umbilical vein to pass from the inferior vena cava through the permeable foramen ovale into the left atrium and systemic circulation [4].

Normally, this membrane separating the AD regresses between 9 and 15 weeks of gestation as the right heart develops, leaving the terminal crest at the top and the Eustachian valve of the inferior vena cava and the Thebesian valve of the coronary sinus at the bottom. During fetal life, the right sinus valve directs oxygenated blood from the inferior vena cava through the foramen ovale to the left side of the heart and into the body. In case of persistence of this valve it results in a separation of the AD into an inlet and an outlet portion. The inflow portion receives systemic venous return from the inferior and superior venae cavae, while the outflow portion contains the right atrial appendage and the tricuspid valve orifice [2]. The dividing membrane in CTD can vary from small to large, can contain fenestrations or be intact, and can obstruct blood flow to the RV [4].

CTD can occur as an isolated cardiac anomaly or be associated with other malformations of right-sided heart structures, such as pulmonary artery stenosis or atresia, tricuspid valve abnormalities, ASD or Ebstein's anomaly [6,7].

Unlike cor triatriatum sinister, which carries a high mortality rate if not intervened, CTD has variable clinical manifestations depending on the degree of division and obstruction of the RA [6].

According to 14 cases reported and collected from 1963 to 2020, the timing and severity of the various clinical manifestations depend mainly on 6 factors in children: The size of the sinoatrial orifices, which determines the degree of obstruction of blood flow from the smooth sinus part of the RA to its cancellous part, which contains the tricuspid valve. The presence of an associated atrial septal defect, the size of which determines the amount of right-to-left shunt. The presence of associated RV hypoplasia or tricuspid valve hypoplasia resulting in limited distensibility and therefore increased right-to-left shunt. The presence of functional RV obstruction created by membrane protrusion through the tricuspid orifice during diastole or the presence of tricuspid valve insufficiency. The presence of associated RV outflow tract obstruction at different levels [5]. In the case presented, the patient had at least 4 of the above factors, such as ASD, RV, tricuspid annulus hypoplasia and alteration in the RV outflow tract.

In patients with severe obstruction, hypoxia is often detected in infancy and corrective surgery should be performed immediately. However, if the CTD membrane is fenestrated or less extensive, the lesion is often recognized only incidentally on echocardiographic imaging, during surgery or autopsy. In these cases, the malformation is asymptomatic and hemodynamically insignificant, and these patients are generally not treated unless they undergo cardiac surgery for other reasons [4, 6]. On the contrary, some patients report palpitations, in-

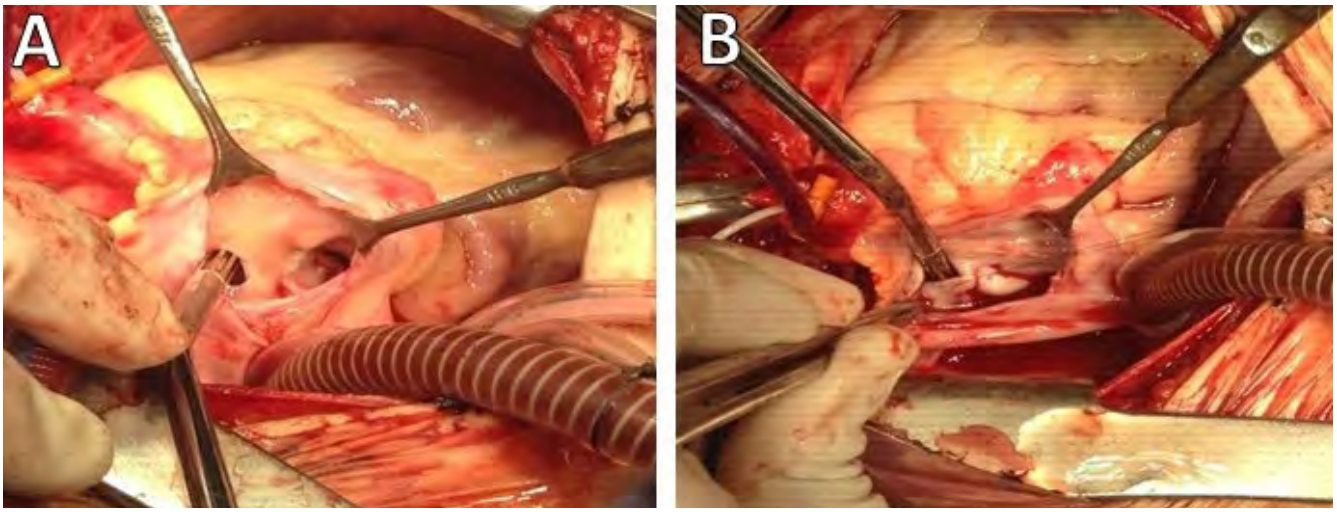

Figure 4. A) There is a hole in the supravalvular membrane of approximately 5 mm. B) The right atrium is observed after membrane resection and direct closure of the atrial septal defect.

creased abdominal circumference reflecting ascites, edema of the pelvic limbs, and on physical examination jugular venous distension and sometimes a systolic heart murmur at the lower sternal border can be observed. In those with a high degree of obstruction and permeable foramen ovale, right-to-left shunting is increased and cyanosis may occur [8].

Diagnosis can be determined by angiography, echocardiography or magnetic resonance imaging [3]. No pathognomonic data on electrocardiogram, chest X-ray or laboratory studies [8].

There are numerous reports on the use of echocardiography as a non-invasive diagnostic tool. However, in a study in which MRI was compared with echocardiography and cardiac angiography in the evaluation of pulmonary venous anomalies, including cases of cor triatriatum (sinister), MRI had a higher detection rate (95%) than the other modalities (69% for angiography and 38% for echocardiography). In general, MRI provides better spatial resolution and superior tissue contrast compared to echocardiography. Another advantage of cardiac MRI is that it can be easily performed in several planes [3].

In the past, the backbone of treatment for symptomatic patients has been surgical resection of the dividing membrane. Recently, percutaneous catheter-based membrane rupture has been reported for obstructive AD membranes and has been suggested as a preferred alternative to open heart surgery, however, definitive treatment of CTD remains surgical, with the current trend towards membrane resection if the patient is symptomatic or if the patient is asymptomatic and undergoing cardiac surgery for other reasons [6,7]. Such is the case of our patient, who benefited from the main procedure (surgical closure of the ASD) to resect the CTD membrane.

In conclusion, CTD is an extremely rare congenital anomaly, with minimal cases reported. There are no reports of satisfactory surgical resolution in Mexico. It can have a variable clinical impact, according to which a therapeutic approach is taken. Surgical resection of the membrane with closure of the interatrial defect remains the preferred treatment in most cases, although percutaneous treatment is still not standardised as a less invasive alternative.

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