Cor Triatriatum: A Tale of Two Membranes

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INTRODUCTION

We report on a case of a neonate with bilateral cor triatriatum with persistent hypoxemia who presented a diagnostic dilemma. Despite extracorporeal membrane oxygenation, multiple echocardiograms, and a cardiac catheterization, the diagnosis was not made until cardiac surgery.

CASE PRESENTATION

A 2-hour-old female neonate born at term via vaginal delivery to a 21-year-old woman with routine obstetric care and a delivery complicated by a tight nuchal cord was transferred to a tertiary neonatology intensive care unit for hypotension and hypoxic respiratory failure. Initial blood pressure was 48/28 mm Hg, and oxygen saturation ranged around 85% on room air. Upon arrival, the patient was intubated and placed on 100% fraction of inspired oxygen and inotropic support. The initial arterial blood gas (fraction of inspired oxygen 1.0) demonstrated pH of 7.42, partial pressure of carbon dioxide 35 mm Hg, and partial pressure of oxygen 53 mm Hg. On physical examination the respiratory rate was 44 breaths/min, heart rate was 105 beats/min, and heart sounds, pulses, and peripheral perfusion were normal. Bedside transthoracic echocardiography demonstrated what was believed to be normal segmental anatomy, a small secundum atrial septal defect with left-to-right shunting, normal systemic and pulmonary venous connections, and normal biventricular systolic function.

With the clinical picture of persistent hypoxemia despite full respiratory support, the patient was started on inhaled nitric oxide and transitioned to an epinephrine infusion for hypotension, without improved clinical status. With persistent hypotension and hypoxemic respiratory failure, partial pressure of oxygen was 40–50 mm Hg, and despite full medical management, the decision was made to place the patient on venovenous extracorporeal membrane oxygenation support. The patient had resolution of hypotension within 24 hours but continued to have low arterial oxygen levels of unclear etiology in the 50–60 mm Hg range. Repeat echocardiographic examinations were performed over the course of 8 days. Although the great artery relationship and systemic and pulmonary venous connections were confirmed normal, the etiology of the persistent hypoxemia was unclear.

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After 9 days of uncomplicated venovenous extracorporeal membrane oxygenation support and persistent hypoxemia, the patient was decannulated and underwent diagnostic cardiac catheterization (Table 1, Figure 1). The angiograms from the femoral venous approach demonstrated flow directed to the right ventricle, but flow also simultaneously filled the left ventricle. The ventriculoarterial connections were normal (Figures 1A and 1B). The combined hemodynamic and angiographic data were concerning for an unusual form of anomalous pulmonary venous return, with the right pulmonary venous flow directed toward the right atrium and the left pulmonary venous flow directed to the inferior vena cava through a confluence above the hepatic veins and the inferior vena cava flow, which was directed through the atrial septum to the mitral valve (Figures 1C and 1D). Unfortunately, no angiograms clearly demonstrated the left pulmonary venous return.

Sedated transesophageal echocardiography performed after catheterization was interpreted as showing total anomalous pulmonary venous return with a misaligned atrial septum, and at this time it was determined to be the cause of the abnormally directed flow from the inferior vena cava to the mitral valve. The following day, repeat transthoracic echocardiography revealed cor triatriatum dexter with a deficient septum primum and abnormal pulmonary venous drainage to the right atrium through a communication in the membrane that separated the superior vena cava and inferior vena cava flow from the tricuspid valve inflow (Videos 1 and 2). A separate membrane extended from the thickened Eustachian valve and connected to what appeared to be the primum septum and divided the left atrium, separating the pulmonary venous return from the mitral inflow, suggestive of cor triatriatum sinister (Videos 3–5).

The patient was referred for surgical repair on day of life 14, and the intraoperative findings were an unusual combination of both cor triatriatum dexter and cor triatriatum sinister, with no true atrial septum. In addition there was separate drainage of a left hepatic vein to the posterior aspect of the right atrium, and the pulmonary venous connections to the left atrium were normal. Specifically, a thick cor triatriatum dexter membrane divided the right atrium, separating the tricuspid valve inflow from the superior and inferior venae cavae. The superior vena cava, inferior vena cava, and a separate leftward hepatic vein all entered the right atrium posterior to this membrane, with blood flow channeled through a small defect that directed the blood flow to the mitral valve. In the left atrium, all four pulmonary veins were separated from the mitral valve inflow by a separate thinner cor triatriatum sinister membrane with a moderate-sized opening superiorly and anteriorly, thus allowing the pulmonary venous flow to be directed toward the tricuspid valve. The relationship of the two membranes to the venous return is illustrated in Figure 2.

The surgical repair consisted of excision and resection of both membranes. A section of untreated autologous pericardium was trimmed to size and used to create an atrial septum, which separated the pulmonary venous return from the systemic venous return. The patient was weaned from cardiopulmonary bypass without difficulty, in normal sinus rhythm with normal hemodynamics, and required no inotropic support. Transesophageal echocardiography demonstrated

Table 1 Hemodynamic data obtained under general anesthesia

Site	Oxygen saturation (%)	Pressure (mm Hg)
SVC	45	
Low SVC	60	
RA	68	a = 12, v = 10, m = 8
High IVC	65	
IVC	65	
MPA	82	32/10, m = 22
RPA	82	32/10, m = 22
LPA	82	32/10, m = 21
RUPV	97	
LUPV	98	
LLPV	97	
LV	66	60/8
Femoral artery	68	55/30, m = 40

IVC, Inferior vena cava; *LLPV*, left lower pulmonary vein; *LPA*, left pulmonary artery; *LUPV*, left upper pulmonary vein; *LV*, left ventricle; *MPA*, middle pulmonary artery; *RA*, right atrium; *RPA*, right pulmonary artery; *RUPV*, right upper pulmonary vein; *SVC*, superior vena cava.
Fraction of inspired oxygen = 0.21.

unobstructed pulmonary venous flow into the left atrium and the mitral valve and unobstructed systemic venous return from the superior vena cava, inferior vena cava, and left-sided hepatic vein into the right atrium and the tricuspid valve. There was no mitral or tricuspid valve insufficiency, and a saline contrast study revealed no residual intracardiac shunting. Biventricular function was normal. The patient was discharged home on postoperative day 4 and continues to do well.

The presence of bilateral cor triatriatum created a physiologic state similar to that of an atrial switch with the systemic venous return directed to the systemic side and the pulmonary venous return directed to the pulmonary side. This is the first reported case of which we are aware of simultaneous cor triatriatum sinister and dexter without a true atrial septum in a cyanotic neonate.

DISCUSSION

Cor triatriatum is a very rare congenital cardiac anomaly with a reported incidence of 0.1% of all congenital heart defects.[1,2] There are two types of cor triatriatum; dexter involves right atrial division, and sinister involves left atrial division. In cor triatriatum dexter, the right atrium is divided into two chambers by exaggerated fetal Eustachian and Thebesian valves, as a remnant from poor regression of the right valve of the right horn of the embryologic sinus venosus, which come together to form an incomplete septum across the lower part of the atrium.[3,4] In cor triatriatum sinister, the left atrium is divided into two chambers by a fibromuscular septum, with the pulmonary veins entering a posterior-superior chamber separated from the anterior-inferior distal chamber containing the mitral valve.[1] The embryologic origin of this defect remains controversial, with two different theories, the malincorporation and entrapment theories.[5]

The presentations of cor triatriatum dexter and cor triatriatum sinister are both related to the degree of obstruction caused by the membranes and the presence of associated defects, which allow the blood to pass through abnormally.[2,3,6,7] Cyanosis is rare in cor triatriatum dexter, but if there is obstruction to the right ventricular outflow tract, cyanosis may be present. Cor triatriatum dexter is often associated with other heart defects, such as pulmonary venous anomalies, tricuspid valve hypoplasia or atresia, and right ventricular hypoplasia. Because of an overall small number of reported cases of cor triatriatum dexter, the exact numbers and associations are not known.[8,9] In cor triatriatum sinister, cyanosis is also rare but can be present on the basis of associated lesions.[2,3] There are multiple reports of cor triatriatum sinister associated with other forms of congenital heart disease in up to 84% of patients, ranging from atrial septal defects and tricuspid regurgitation to complex heart disease, including transposition of the great arteries and tetralogy of Fallot.[2] Obstructive cor triatriatum sinister usually presents in infancy; however, there are multiple case reports of cor triatriatum sinister presenting in adults with chronic atrial fibrillation. The Mayo Clinic reported a series of 35 patients with cor triatriatum sinister in which the mean age at the time of diagnosis was 27.4 years.[2,6,7]

The diagnosis of cor triatriatum dexter and cor triatriatum sinister is usually made by echocardiography.[2,5,9] It is important to differentiate the cor triatriatum sinister membrane from a supravalvar mitral ring, and the left atrial appendage is used as an anatomic landmark. The cor triatriatum sinister membrane is seen above the left atrial appendage, and almost always the pulmonary veins drain into the superior chamber. The supravalvar mitral ring is seen below the left atrial appendage and just above the mitral valve annulus. Angiography has been largely replaced by echocardiography as a diagnostic tool in both conditions; however, when the diagnosis is unclear, the next step is to perform cardiac catheterization to obtain hemodynamics and angiography.[2-4] Alphonso *et al.*[1] reported a series of 28 patients in which 32% with atypical anatomy of cor triatriatum sinister required cardiac catheterization for full demonstration of the defects, compared with all patients with classic cor triatriatum sinister anatomy who were diagnosed by echocardiography alone. Additionally, a case report by Edwin *et al.*[7] describes a patient with cor triatriatum sinister in the absence of the interatrial septum, which was diagnosed in the operating room, illustrating the limitations of echocardiography in the setting of cor triatriatum with unusual and more complex anatomy.

Definitive treatment for cor triatriatum dexter is surgical, with the current trend for resection of cor triatriatum dexter if the patient is symptomatic or if asymptomatic and undergoing cardiac surgery for other reasons.[3] Treatment for cor triatriatum sinister is surgical resection, with good results and low risk for recurrence.[2] The overall mortality and morbidity for cor triatriatum sinister are low, with reports of survival of 96% at 5 years and 88% at 15 years, and low risk for reoperation.[1,2]

Reports of bilateral cor triatriatum are limited. One reported case by Al-Mousily *et al.*[4] described an 11-month-old female infant with bilaterally divided atrial chambers and associated right upper pulmonary vein stenosis, successfully treated with surgical resection of both membranes.

When a neonate remains persistently cyanotic despite extracorporeal membrane oxygenation support, rare and unusual cardiac anatomic variations should be considered. A detailed histopathologic description of the surgical specimen is not available, and therefore, there could be no correlation between anatomic findings by imaging and intraoperative inspection. In our case, despite multiple echocardiographic studies and cardiac catheterization, the anatomy could be clearly delineated only by direct intraoperative inspection.

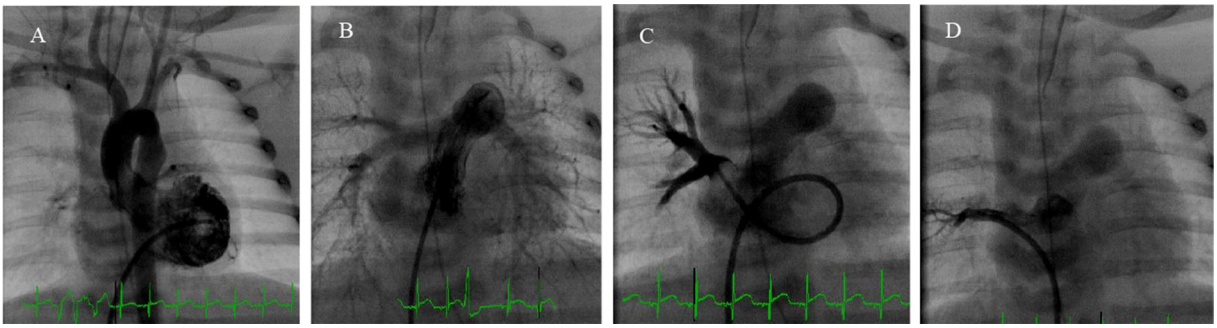

Figure 1 Angiograms. **(A)** Left ventricular angiogram from femoral venous approach showing direct connection of systemic veins to the left ventricle, which is in continuity with the aorta. **(B)** Right ventricular angiogram via femoral venous approach demonstrating normal systemic venous connection to main pulmonary artery. **(C)** Right upper pulmonary vein angiogram via femoral venous approach demonstrating apparent connection to right atrium. **(D)** Right lower pulmonary vein angiogram via femoral venous approach demonstrating apparent connection to right atrium.

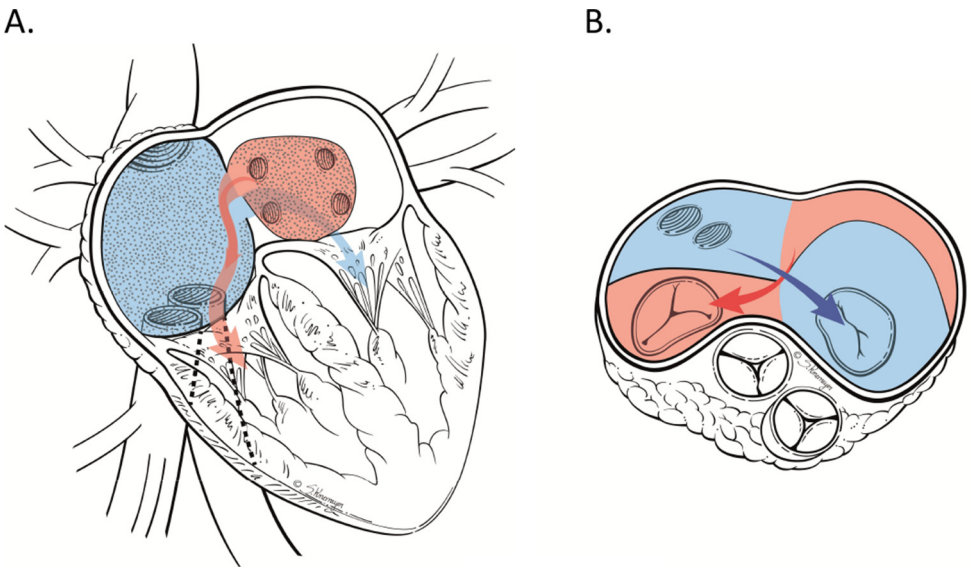

Figure 2 Intraoperative anatomy. **(A)** Four-chamber view of the heart in situ from anterior looking posterior with cor triatriatum dexter membrane in *blue* and cor triatriatum sinister membrane in *red*. **(B)** En face view looking down at the atrium with the membranes in relation to each other and the atrioventricular valves.

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SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2018.08.003.

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