Cor triatriatum Dexter: A Rare Cause of Intermittent Cyanosis in the Newborn

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ABSTRACT

We present the case of an infant who was prenatally diagnosed with a size discrepancy between the right and left ventricles, but postnatally was found to have cor triatriatum dexter. The patient was initially noted by fetal echocardiogram to have a small right heart. Prenatally, the right ventricle appeared smaller than the left ventricle, although it was considered in the low normal range for size at that time. Nevertheless, because of this discrepancy, a postnatal echocardiogram was obtained and revealed normal biventricular size and systolic function. Surprisingly, a large, mobile membrane was found above the tricuspid valve, without restriction of flow across the tricuspid valve. It was noted to traverse across the tricuspid valve during diastole and regress back into the atrium during systole without restriction of flow across the tricuspid valve. An atrial septal defect was also present (Figure 1). Chest x-ray revealed a normal cardiothymic silhouette and clear lungs.

Case Report

A one day old infant was born full term via an uncomplicated, normal spontaneous vaginal delivery with a birth weight of 3.27 kilograms and Apgar scores of 9 at 1 minute and 9 at 5 minutes. Prenatally, the right ventricle appeared smaller than the left ventricle, although it was considered in the low normal range for size at that time. Nevertheless, because of this discrepancy, a postnatal echocardiogram was obtained and revealed normal biventricular size and systolic function. Surprisingly, a large, mobile membrane was found above the tricuspid valve, without restriction of flow across the tricuspid valve. An atrial septal defect was also present (Figure 1). Chest x-ray revealed a normal cardiothymic silhouette and clear lungs.
A repeat echocardiogram on day of life three confirmed a mobile membrane superior to the tricuspid valve without restriction of blood flow across the valve, as well as an atrial septal defect with right to left flow. Electrocardiogram was normal with sinus rhythm.

The infant was monitored in the neonatal intensive care unit where she remained clinically well and was eventually discharged home with a pulse oximeter and oxygen as needed. At home, she was initially well with no tachypnea, feeding intolerance or persistent cyanosis, but with time her parents reported a steady decline in her baseline saturations. By two weeks of life, the baseline saturation had reached the low 80s, which prompted surgical removal of the cor triatriatum membrane. Intraoperative findings demonstrated that the membrane spanned from the outlet of the superior vena cava down to the Eustachian valve and over to the septal leaflet of the tricuspid valve. While there was no obstruction at the level of the tricuspid valve, the cor membrane preferentially shunted blood flow right to left across an 8 mm secundum atrial septal defect. The membrane was removed and the atrial septal defect was closed primarily. Postoperative transesophageal echocardiogram demonstrated no residual membrane, no residual interatrial shunt, normal inferior and superior vena cava flow, normal left and right ventricular size and function, and normal tricuspid valve. After an uncomplicated postoperative course, the patient was discharged home on postoperative day three with normal saturations and no oxygen requirement.

Discussion

During fetal life, the function of the Eustachian valve of the sinus venosus is to divert oxygenated blood from the inferior vena cava to the left atrium via the foramen ovale. With development of the heart, the sinus venosus is typically incorporated into the right atrium. Cor triatriatum dexter is a rare entity in which persistence of the right-sided valve creates a division of the right atrium [4]. Often, patients are asymptomatic because the membrane is fenestrated or small enough that systemic venous flow passes unobstructed to the outflow chamber. If complications requiring surgical intervention do occur, they are usually secondary to right sided obstruction causing signs and symptoms of heart failure or intractable arrhythmia [5]. In addition, one report describes the case of a 22 year old woman who suffered from a myocardial infarction and pulmonary embolism thought to be secondary to a thrombus found on the thick and calcified ridge within the right atrium [6].

For our patient, the cor membrane preferentially diverted deoxygenated blood through a secundum atrial septal defect, resulting in intermittent desaturations that evolved to persistent cyanosis. This is important to keep in mind in the context of a lack of tricuspid valve obstruction or clinical signs of heart failure. León et al. [7] described a similar case of a newborn with a persistent Eustachian valve, but the Eustachian valve is embryologically different and a much more common finding than cor triatriatum dexter. Also, that patient did not require surgical intervention. While there is one reported case of persistent mild cyanosis associated with cor triatriatum dexter and a secundum atrial septal defect in a nine year old [8], our patient represents the first case of cor triatriatum dexter presenting as neonatal cyanosis without signs of heart failure. We suspect the cor membrane may have been difficult to identify prenatally given its lack of rigidity within the atrial chamber. Additionally, the borderline small right heart structures demonstrated on fetal echocardiogram were likely secondary to the decreased flow across the tricuspid valve due to the cor membrane. Although rare, cor triatriatum dexter should be considered in both the differential for fetal echocardiograms that suggest small right sided structures, as well as neonatal cyanosis with or without accompanying signs or symptoms of heart failure.

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Conflicts of Interest

The authors have no financial or non-financial conflicts of interest to disclose. No affiliations or any personal, racial and intellectual properties to disclose.

Contributor’s Statement

Dr. Ryan Serrano drafted the initial manuscript and approved the final manuscript as submitted.

Dr. Brandy Hattendorf reviewed and revised the manuscript, assisted with image collection, and approved the final manuscript as submitted.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the institutional committee.

References


