



## Venous-Arterial ECMO as a Vital Bridge for Survival in a Neonate with Cor-triatritium Dexter

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### Abstract

Cor-triatritium dexter is an extremely rare congenital heart disease which can present with varied clinical manifestations. These range from being asymptomatic, incidentally diagnosed during cardiac surgery, right-sided heart failure, cyanosis and death in the neonatal period. This heart defect results from remnants of the right valve of the sinus venosus causing varying degrees of obstruction to the tricuspid valve in flow. We present a neonate with cor-triatritium dexter who initially failed a car seat tolerance screening test and rapidly deteriorated into cardiogenic shock with tricuspid valve inflow occlusion after closure of Patent Ductus Arteriosus (PDA). The PDA was completely unresponsive to prostaglandins, necessitating emergent institution of venous-arterial extra corporeal membrane oxygenation (VA-ECMO), which provided a vital bridge for survival followed by early primary surgical resection of the cor-triatritium dexter membrane. Without VA-ECMO, an adverse outcome may have ensued from a surgically correctable congenital heart defect. In this report, we stress both the need for a high index of suspicion of this rare anomaly and the value of VA-ECMO as a bridge to successful surgical correction in a symptomatic neonate with a cardiac defect that offers excellent prognosis post-surgical correction.

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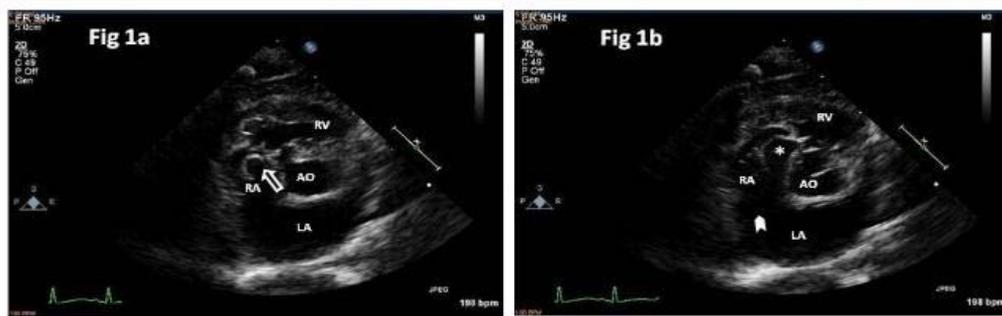
**Keywords:** Cor-triatritium dexter; VA-ECMO; Neonate

### Introduction

Cor-triatritium dexter is an extremely rare congenital heart defect with prevalence of less than 0.01% [1]. This results from the complete persistence of the right valve of the embryonic sinus venosus, also known as the Eustachian valve, as a membrane or fibro muscular band [2]. This results in the division of the right atrial chamber into two compartments. The membrane protrudes into the right ventricular chamber through the tricuspid valve, causing choking of the right ventricular inlet and altered hemodynamics. This allows obligatory right to left shunting at the atrial septal level resulting in profound cyanosis. Herein we report a case of newborn that failed a car seat tolerance (CST) screening test that was successfully treated by primary cardiac surgical repair after a brief course of Venous-Arterial Extra Corporeal Membrane Oxygenator (VA-ECMO) at University of Maryland Medical Center (UMMC). Uniquely, this is the first reported case emphasizing the importance of VA-ECMO to serve as a vital bridge for survival in a neonate.

### Case Presentation

A female infant was born at 36 6/7 weeks of gestation via spontaneous vaginal delivery to a 20 year old mother at an outside hospital. There was normal prenatal care and no underlying family history of congenital heart disease. All routine maternal prenatal screening labs were negative. Apgar scores of the newborn at 1 and 5 minutes were 8 and 9, with birth weight of 2665 g. The baby was transferred to the newborn nursery soon after birth and was bottle fed formula with volumes ranging 15-30 ml for approximately 24 hrs. As part of routine discharge planning of any infant born prior to 37 weeks gestation, a CST screening test was performed close to 41 hrs of life. Once placed in the car seat, the baby began having episodes of oxygen de-saturations (65%-88%) while breathing room air. Failing the test, the infant was quickly transferred to the radiant warmer for respiratory support. Due to continued de-saturations, the baby was placed on 100% oxygen delivered through a 2 L high flow nasal cannula with minimal change in oxygen saturations. She was immediately transferred to the newborn intensive care unit (NICU) at UMMC to rule out cyanotic congenital heart disease.



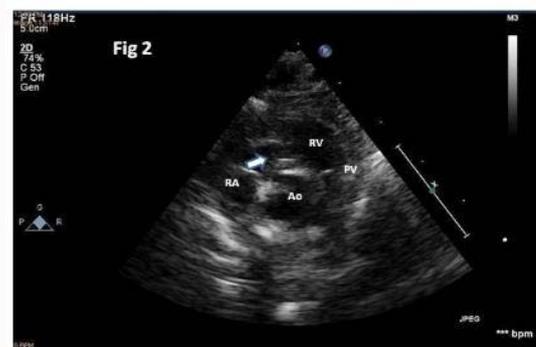
**Figure 1:** (A) Parasternal short axis echocardiographic image clip taken in atrial diastole demonstrating the mobile “cor” membrane just above the tricuspid valve inlet (hollow arrow).

(B) Clip taken during atrial systole demonstrating the cor membrane flopping dynamically into the inflow path of the tricuspid valve causing obstruction (asterisk). Additionally, a large atrial septal defect is visible (solid white arrow). RV: Right Ventricle; RA: Right Atrium; AO: Aorta; LA: Left Atrium

Following admission to the NICU, the newborn was found to have central cyanosis with oxygen saturations between 60%-75% despite remaining on 100% oxygen through 4L of high flow nasal cannula. The physical exam was otherwise remarkable for a baby with vigorous cry with no signs of respiratory distress. Cardiovascular examination revealed a quiet precordium with normal heart sounds and no audible cardiac murmur. Chest x-ray showed pulmonary oligemia with normal lung volumes and normal sized heart. Arterial blood gas confirmed severe hypoxemia with partial pressure of oxygen at 35mmHg on 100% oxygen.

The baby was electively intubated in the NICU and placed on the high frequency oscillator in an attempt to improve oxygenation. Simultaneously, pediatric cardiology was consulted for persistent neonatal cyanosis. Initial echocardiogram demonstrated a redundant “cor” membrane in the right atrial chamber just above the tricuspid valve inlet in atrial diastole (Figure 1A) flopping into the orifice of the tricuspid valve during atrial systole (Figure 1B). A small hole in the membrane was visible through which some forward blood flow passed into the pulmonary arteries. A hypertrophied small cavity right ventricle, right to left shunt across the large atrial septal defect, and right to left bowing of the atrial septum were also present. Also, remarkably, a patent ductus arteriosus (PDA) could not be visualized. The baby was immediately started on a continuous prostaglandin (PGE1) infusion in addition to a dopamine infusion and hydrocortisone in an effort to improve systemic oxygenation. However, the infant continued to deteriorate progressively to respiratory failure from cardiogenic shock secondary to tricuspid inflow blockage and worsening right to left inter-atrial shunting. In an effort to open the PDA, an intravenous bolus of PGE1 (180 micrograms) was attempted with no response. Bedside echocardiogram performed serially for several minutes after the PGE1 infusion and bolus demonstrated no ductal flow on color Doppler. At this point, the baby was critically ill with oxygenation index exceeding 40 and the ECMO team was activated. VA-ECMO cannulation was performed at the bedside at a point approximately 3 hours after her arrival to the NICU. Arterial blood gases immediately improved and baby was transferred to the cardiac section of the pediatric intensive care unit (PICU) for further cardiac management.

Repeat echocardiogram after institution of VA-ECMO interestingly showed that the right jugular venous cannula had deflected the cor-triatriatum dexter membrane away from the tricuspid valve inlet and pinned it against the Inferior Vena Cava-Right Atrial junction (Figure 2). ECMO flows were however adequate and the infant stabilized thereafter. There was some concern



**Figure 2:** Echocardiographic image clip taken in parasternal short axis view after emergent institution of Venous-Arterial ECMO from the right neck showing how the venous cannula had pinned the “cor” membrane away from the tricuspid valve inlet thereby allowing temporary unobstructed opening of the tricuspid inlet (solid arrow).

RV: Right Ventricle; RA: Right Atrium; AO: Aorta; PV: Pulmonary Vein

regarding adequate size of the right ventricle. However, the tricuspid valve annulus diameter z-score was reassuring at -2.0. The following morning, the baby was taken to the operating room. Intra-operative findings were consistent with the diagnosis of cor-triatriatum dexter. The right atrial membrane that was resected was found to be pedunculated and attached to the crista terminalis, eustachian valve, and tricuspid valve annulus. The atrial septal defect was closed primarily. Her post-operative course was fairly uncomplicated. She had a small residual atrial level communication, and was diagnosed with unilateral partial paralysis of the vocal cord that spontaneously resolved. She was evaluated by a pediatric geneticist and was found to have a normal single nucleotide polymorphism array. The baby was discharged on hospital day 13 on low dose aspirin. She continues to gain weight, is currently thriving well and has regular follow up appointments with pediatric cardiology.

## Discussion

This is the first reported case emphasizing the importance of VA-ECMO in a neonate to serve as a vital bridge for survival prior to successful treatment by primary cardiac surgical repair for cor triatriatum dexter. Initiation of VA-ECMO was of vital importance in this case due to the rapid development of cardiogenic shock from presumed early PDA closure. Although only a few reports of cor-triatriatum dexter that lead to cyanosis with an otherwise normal exam in a new born exist in the literature [3,4] our case is unique

in that closure of the PDA was refractory to PGE1 and precipitated rapid deterioration into cardiogenic shock with profound hypoxemia and respiratory failure.

When diagnosing cor-triatritium dexter, the physical exam is rarely helpful, as a murmur may not be audible due to lack of turbulent blood flow across the membrane. The chest X-ray usually demonstrates a normal cardiac silhouette and normal lung volumes. Intermittent episodes of central cyanosis and de-saturations can be missed in the immediate newborn period. Clinical presentation can range from being asymptomatic in the newborn period to reports of pulmonary hypertension and cardiac failure at an older age [2,5-8]. Thorough echocardiographic imaging is therefore mandatory. Both short term and long term prognosis is usually excellent following surgical repair [9]. The use of ECMO as a life-saving measure for a cyanotic neonate with isolated cor-triatritium dexter has been reported in older children [10], but none have been previously reported in a neonate. While limited case series on symptomatic neonatal presentations can be found, none of the neonates discussed progressed to such extremis as the case reported herein. A 4 month old infant with cor-triatritium dexter required ECMO secondary to pulmonary hypertension, but had associated structural heart defects including pulmonary vein stenosis, large ventricular septal defect and atrial septal defect. In that case, the infant was decannulated, but died prior to surgical repair [11].

Additionally, herein, we present a case of a newborn with cor-triatritium dexter who failed a car seat test. CST screening test is a common pre-discharge assessment requisite for newborns born <37 weeks gestational age or <2500 g. The utility of the test remains in identifying infants at risk for adverse cardiopulmonary outcomes after discharge from the newborn nursery [12,13]. In this neonate, the CST screening test, offered the opportunity to screen an otherwise healthy-appearing baby for this potentially lethal congenital heart defect. If this screening had not been performed, an adverse outcome may have ensued. This raises very important questions about the babies who are deemed eligible for the CST screening test. It also raises the spectre of the importance that all newborn nurseries, even low risk newborn units, adopt routine neonatal pulse oximetry screening that may serve as an important backup [14].

## Conclusion

High index of suspicion is necessary to diagnose rare conditions such as cor-triatritium dexter in an otherwise normal appearing neonate with repeated desaturations and cyanosis in the immediate newborn period. Our case report provides emphasis of the importance of VA-ECMO as a vital bridge to survival in a neonate until primary cardiac repair despite failed attempts to pharmacologically open the closed PDA.

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