Cortriatriatum Dexter: A Rare Disease and Easily Missed Diagnosis

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Abstract
Cortriatriatum dexter is an extremely rare congenital heart anomaly in newborns characterized by anatomic division of the right atrium by a membrane which divides the right atrium into three chambers. A case of cortriatriatum dexter is reported in a 38-week GA infant presenting with respiratory distress and cyanosis. She was initially diagnosed with persistent pulmonary hypertension and eventually needed extra corporeal membrane oxygenation (ECMO) therapy. Cortriatriatum dexter was later diagnosed before discontinuing the ECMO. The membrane removal was successfully performed at day seven of life without complications.

Keywords: Cortriatriatum dexter; Newborn; Rare disease

Abbreviations

Introduction
Cortriatriatum dexter (CTD) is the remnants of fetal right sinus valve which usually regresses between 9 and 15 weeks of gestation. It is an extremely rare condition, accounting for 0.1% of congenital heart disease [1]. CTD characterized by anatomic division of the right atrium (RA) by a membrane, which divides the RA into two chambers [2]. Clinical manifestations depends on the severity of obstructed blood flow and the amount of desaturated blood across the atrium septum [3], ranging from asymptomatic to respiratory distress and cyanosis. If the obstruction of blood flow is mild, it is sometimes incidentally discovered by autopsy or echocardiogram [4]. Importantly, it might be associated with other congenital heart defects such as pulmonary artery stenosis or tricuspid valve atresia [5]. A case of CTD is reported in infant presenting with respiratory distress and cyanosis. The diagnosis was missed due to initial persistent pulmonary hypertension (PPHN).

Case Presentation
A 38-week-GA female infant, birth weight of 3.750 grams was born by cesarean section to a 34-year-old G1P0 woman with an uneventful pregnancy. The Apgar scores were 8 and 9 at 1 and 5 minutes. Four hours after birth, the infant developed tachypnea and cyanosis; oxygen box 10 L/min and antibiotics were given then oxygen saturation (SpO2) increased to 99-100%. At 36 hours of life, she developed severe respiratory distress, labile SpO2 while crying with a marked drop in SpO2 to 80–90%, and differential cyanosis. Physical examination revealed normal heart sounds without heart murmur and equal breath sounds. Sepsis work-up was negative and chest radiograph was normal. Echocardiogram demonstrated right-to-left shunt across patent foramen ovale (PFO), mild tricuspid regurgitation with tricuspid regurgitation peak gradient (TRPG) of 33 mmHg, mild left ventricle (LV) D-shape, and suspected hyperechoic mass size 4 × 6 mm at lateral border of RA. She was diagnosed with persistent pulmonary hypertension (PPHN) and was intubated and placed on high frequency oscillatory ventilation (HFOV), sildenafil, iloprost, dopamine, and adrenaline, then referred to our hospital.

On day three of life, despite HFOV with fraction of inspired oxygen (FiO2) of 1.0, inhaled nitric oxide, iloprost, adrenaline, dopamine, norepinephrine, and milrinone, her SpO2 remained under 85%. Echocardiogram revealed a 3-millimeter flapped PFO 3 mm with right-to-left shunt, closing patent ductus arteriosus (PDA), suspected hyperechoic mass of 5 mm in diameter at lateral wall of RA, small right ventricle, mild RA enlargement with interatrial septal deviation. Eventually, she was placed on venous-arterial extracorporeal membrane oxygenation (VA-ECMO) for hemodynamic support.

During full ECMO support, clinical condition improved dramatically with no respiratory distress and SpO2 could be maintained above 95%. During weaning from ECMO, on day six of life, she developed significant desaturation. The third echocardiogram revealed a thin redundant membrane in the lower part of RA below the coronary sinus orifice and just above the TV with significantly obstructed flow across the membrane opening causing coronary sinus dilatation, 5 mm flap PFO with deviation of the atrial septum to the left atrium (LA) and markedly right-to-left shunt, RA, LA and LV enlargement, left ventricular hypertrophy, and mild mitral valve regurgitation (Figure 1). The infant was then sent for surgery and surgical findings confirmed the diagnosis of CTD (Figure 2). This membrane nearly obstructed total systemic venous blood flow draining into the RV. Membrane resection and atrial septal defect closure were performed. After the operation, she was extubated on day 12 of life and discharged on day 30 of life. Her growth and development at follow-up clinic were normal. Latest echocardiogram showed no residual heart defects.

Discussion
This case was initially diagnosed as PPHN due to persistent cyanosis, labile SpO2 that could reach 100% when systemic blood pressure was at a high level and evidence of right-to-left shunt across PFO by echocardiogram. This case might also have had PPHN on the first few days because there was differential cyanosis indicating right-to-left shunt across PDA. CTD was eventually diagnosed by the third time echocardiogram. Severe cyanosis could be explained by nearly total obstruction of blood flow from RA to RV by the membrane with a tiny hole, diverting most of the venous...
Multiple echocardiographic views are warranted [3]. In most cases, more than one diagnostic imaging modality is necessary to reach a final conclusion because each modality has its own limitation [8].

Management of CTD depends on the degree of blood flow obstruction to RV ranging from nonetosurgical resection. The short and long-term prognoses are generally good [9]. In this case, the patient had severe cyanosis and required surgery. Despite percutaneous catheter disruption having been reported [10], surgery is the preferred choice of treatment.

In conclusion, the diagnosis of CTD requires a high degree of suspicion, especially in a newborn with persistent cyanosis unexplained by other common causes. Multiple echocardiographic views are warranted [3]. In most cases, more than one diagnostic imaging modality is necessary to reach a final conclusion because each modality has its own limitation [8].

Figure 1: Transthoracic echocardiographic image in subcostal short axis view illustrates a membrane in RA lines above tricuspid valve, (a); four-chamber view shows minimal blood flow across membrane through tricuspid valve, (b). LA: Left Atrium; LV: Left Ventricle; RA: Right Atrium; RV: Right Ventricle.

Figure 2: Intraoperative findings: a photograph of CTD membrane in RA before surgical resection, (a); after membrane resection at divided area, the fossa ovalis is shown, (b); a piece of the CTD membrane, (c). CTD: Cortriatriatum Dexter, FO: Fossa Ovalis, M: Membrane; RA: Right Atrium; RV: Right Ventricle.
and unresponsiveness to oxygen supplement. The membrane is elusive and needs careful evaluation. Prompt surgical treatment is mandatorily indicated in those with severe cyanosis.

**Disclosure Statement**

No conflict of interest to declare.

**References**


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