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CASE REPORT

A Case Series of Patients with Cor Triatriatum Dexter: Unique Cause of Neonatal Cyanosis

Chad A. Mackman · Jennifer L. Liedel · Ronald K. Woods · Margaret M. Samyn

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Abstract Cor triatriatum dexter is a rare congenital heart defect that can lead to cyanosis in a newborn with an otherwise normal exam. The initial evaluation of these patients typically focuses on searching for a pulmonary etiology for arterial desaturation, which often leads to a negative work up. When cardiac evaluation is performed, it may be challenging because the heart lesion can be difficult to visualize on an echocardiogram. The diagnosis requires a high index of suspicion and thorough echocardiographic imaging. Once diagnosed, surgical repair can alleviate the shunt created by the defect. This case series describes all patients (3) with cor triatriatum dexter seen at Children's Hospital of Wisconsin from 2000 to 2013.

Keywords Cor triatriatum dexter · Neonatal cyanosis · Right atrial membrane · Congenital heart disease

C. A. Mackman · M. M. Samyn
Division of Cardiology, Department of Pediatrics, Medical College of Wisconsin, Milwaukee, WI, USA

C. A. Mackman (✉) · J. L. Liedel · R. K. Woods · M. M. Samyn
Children's Hospital of Wisconsin, 9000 W Wisconsin Ave MS 713, Milwaukee, WI 53226, USA
e-mail: cmackman@chw.org

J. L. Liedel
Divisions of Neonatology and Critical Care, Department of Pediatrics, Medical College of Wisconsin, Milwaukee, WI, USA

R. K. Woods
Division of Cardiothoracic Surgery, Department of Surgery, Medical College of Wisconsin, Milwaukee, WI, USA

Abbreviations

ASD	Atrial septal defect
CHW	Children's Hospital of Wisconsin
DOL	Day of life
LA	Left atrium
PDA	Patent ductus arteriosus
PFO	Patent foramen ovale
RA	Right atrium or right atrial
RV	Right ventricular
TV	Tricuspid valve

Cases

Case 1: A 3,200 g term female infant (with a negative septic workup, normal physical exam, and normal chest radiograph) developed intermittent arterial oxygen desaturation (70–80 %) leading to intubation on day of life (DOL) 4. The initial echocardiogram was concerning for a ventricular septal defect (VSD), patent foramen ovale (PFO), and a mildly dysplastic tricuspid valve (TV), which would not be anticipated to cause cyanosis. Upon transfer to Children's Hospital of Wisconsin (CHW), a repeat echocardiogram showed a membrane in the right atrium (RA) consistent with cor triatriatum dexter. Right to left shunting across the PFO was seen, as was evidence of elevated RA pressures due to the membrane's partial obstruction of TV inflow. The TV appeared structurally normal, and the ventricular septum was intact (Fig. 1).

Surgical inspection confirmed a membrane with attachment rightward and posterior to the crista terminalis and a line of attachment extending to the septal region of the TV annulus. The caval inflow and small atrial septal

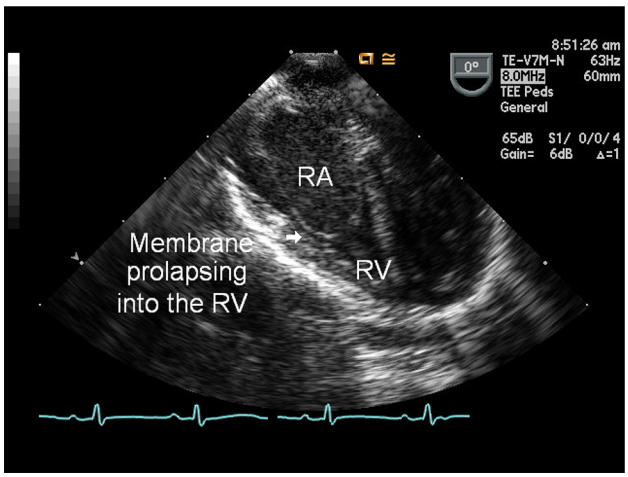

Fig. 1 Transesophogeal echocardiographic image shows the cor triatriatum dexter membrane prolapsing through the TV into the right ventricle (RV)

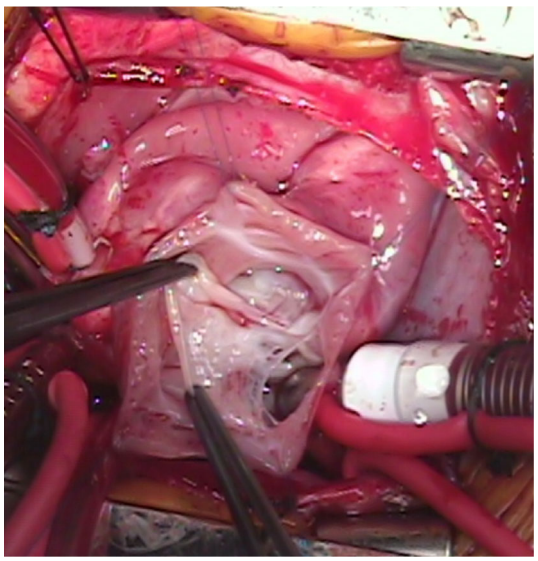

Fig. 2 Intra operative view, looking down toward the TV, shows the RA membrane held by forceps

defect (ASD) were proximal (above) to the membrane (Fig. 2). The membrane was resected, and the ASD was closed. Her post-operative recovery was uncomplicated, and she was discharged home 6 days after surgery. She had no oxygen requirement and no complications noted in follow-up.

Case 2: A 3,200 g term female infant had an arterial oxygen saturation of 73 % on DOL 1. Chest X-ray and physical exam revealed no abnormalities. An echocardiogram demonstrated a thin, freely mobile membrane within the RA, consistent with cor triatriatum dexter. The membrane directed the systemic venous blood through the PFO into the left atrium (LA). It also prolapsed through the TV, obstructing right ventricular (RV) inflow. Her oxygen saturations fell into the 60 s on DOL 2, necessitating prostaglandin therapy for ductal patency to increase pulmonary blood flow. This improved oxygen saturations to the low 80 s.

On DOL 8, she underwent repair with resection of the membrane, suture closure of the PFO, and patent ductus arteriosus (PDA) ligation. She was discharged to home on room air on DOL 13. She had no complications noted in follow-up.

Case 3: A 1,590 g female infant was born at 36 weeks' gestational age after a prenatal course complicated by intrauterine growth restriction. On exam she was noted to have microcephaly and abnormal facies. Because of concern for a genetic syndrome, an echocardiogram was obtained, suggesting a structurally normal heart with a PFO. She was transferred to CHW on DOL 15 for genetic evaluation and care of her feeding difficulties.

On arrival she had room air arterial oxygen saturations in the high 90 s with transient desaturation to the 70 s. Her exam revealed mild micrognathia, synophrys, and low set ears. She had a grade 1–2/6 systolic ejection murmur at the left midsternal boarder radiating to bilateral upper sternal borders. Finding no pulmonary cause for her desaturation, a repeat echocardiogram was performed. A redundant fenestrated membrane above the TV, consistent with cor triatriatum dexter, was identified (Fig. 3) with bidirectional flow across a PFO. The TV was structurally normal; however, the RV cavity was mildly hypoplastic, as was the pulmonary valve (Z score -3.9). She was diagnosed with Cornelia de Lange Syndrome and discharged home at 1 month of age without need for supplemental oxygen. In follow-up, she developed persistent desaturation (low 90 s). Follow-up echocardiogram showed desaturated systemic venous return streaming preferentially toward the LA through the PFO due to the presence of cor triatriatum dexter. This RA membrane also created progressive obstruction to TV inflow. At 6 months old, she underwent resection of the membrane and PFO closure. Her post-operative course was complicated by prolonged pericardial effusion, resolving with medical therapy. Within 3 months of surgery, she was off oxygen and all cardiac medications with no significant residual complications.

Discussion

Cor triatriatum dexter is a rare heart defect which accounts for about 0.1 % of all congenital heart malformations [9]. During embryologic development, the right valve of the sinus venosus extends across the RA. Regression of this structure results in the crista terminalis, Eustachian valve, and Thebesian valve [4]. When this tissue forms a network of

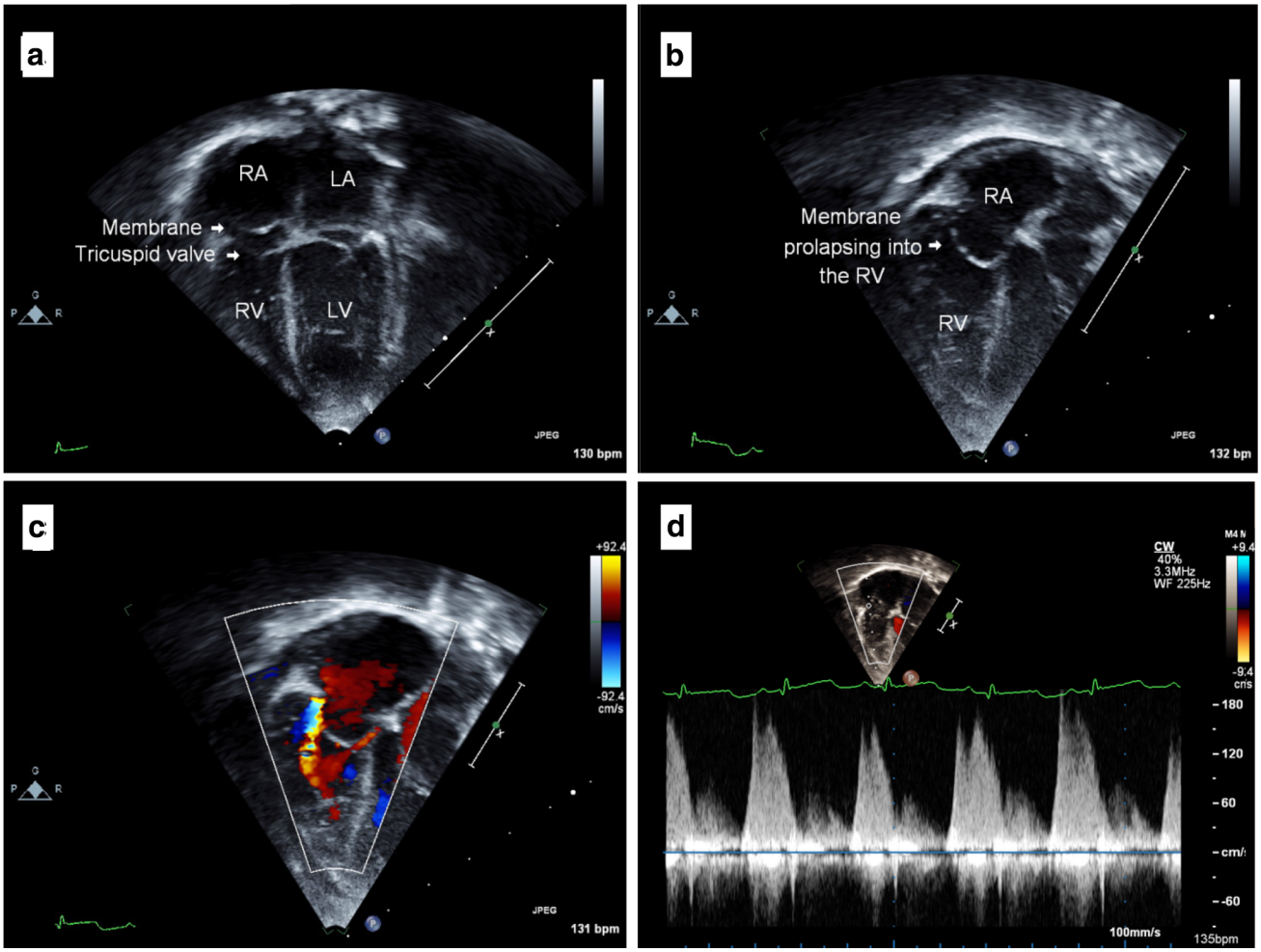

Fig. 3 **a** Echocardiographic apical 4 chamber image during systole shows cor triatriatum dexter with the membrane positioned in the right atrium above the tricuspid valve. **b** Echocardiographic apical 4 chamber image during diastole shows the membrane prolapsing through the tricuspid valve into the RV. **c** Echocardiographic apical 4 chamber image with *color* Doppler shows turbulent blood flow across the membrane suggesting obstruction to RV inflow. **d** Echocardiographic apical 4 chamber image with spectral Doppler shows elevated blood flow velocity across the membrane with an elevated mean gradient (7 mmHg) consistent with inflow obstruction

fenestrated fibers, it is a Chiari's network [2]. When these fibers form a membrane, it is diagnosed as cor triatriatum dexter.

The clinical significance of cor triatriatum dexter depends on the degree of obstruction to blood flow through the TV and the amount of streaming of desaturated blood across the atrial septum. Mild or no obstruction leads to an asymptomatic patient, with the membrane discovered on autopsy (after death from unrelated cause) or as incidental finding on echocardiogram performed for unrelated reasons [3, 7]. More severe obstruction leads to poor RV filling and compromised pulmonary blood flow, as seen in case 2. Some membranes can cause streaming of the blood from the RA across the atrial septum to the LA, as seen in our cases. These patients have mild cyanosis from shunting but no evidence of heart failure.

The diagnosis of cor triatriatum dexter can be difficult. Intermittent or mild arterial oxygen desaturation is suggestive of other more common causes such as transient tachypnea of the newborn, persistent pulmonary hypertension, pneumonia, or sepsis. This is seen in case 1, where a pulmonary process was suspected and led to escalation of respiratory support.

The physical exam is rarely helpful when diagnosing cor triatriatum dexter. The membrane rarely causes enough turbulent blood flow to create an audible murmur. Blood gas analysis may also be of little benefit. Chest X-ray usually demonstrates a normal cardiac silhouette with normal pulmonary vascular markings.

On echocardiogram, the thin membrane can elude detection, as in cases 1 and 3. The diagnosis of cor triatriatum dexter should be considered in a patient with arterial desaturation unresponsive to oxygen when alternative causes have been excluded. Echocardiographic evaluation of the RA from multiple windows is necessary to identify this membrane, and repeat echocardiogram at a

tertiary referral center may be needed. In addition, MRI and 3D echocardiogram are viable imaging modalities for making this unusual diagnosis but are not usually required [1, 5, 6].

Management of cor triatriatum dexter varies based on the severity of RA obstruction. With limited or no obstruction, no intervention is required. If there is obstruction to RV inflow or development of cyanosis due to shunting across the atrial septum, the RA membrane can be resected, and the atrial septum can be closed. Percutaneous catheter-based intervention has been described [8], but a surgical approach is curative and does not usually require re-intervention.

Conclusion

Cor triatriatum dexter is a rare congenital heart defect to be considered in any neonate with unexplained cyanosis. The diagnosis can be challenging, so careful evaluation of the RA by echocardiogram is essential to recognizing this lesion.

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