Cor triatriatum dexter: A rare cause of cyanosis during neonatal period

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ABSTRACT

Cor-triatriatum dexter is an extremely rare congenital heart defect in which there is complete persistence of the right valve of embryonic sinus venosus that results in partitioning of the right atrium into a smooth and trabeculated portion. The smooth portion receives venous blood from inferior vena cava, superior vena cava, and coronary sinus while the trabeculated portion contains the right atrial appendage and the opening of tricuspid valve.

We report a 1-week-old child who presented with intermittent episodes of central cyanosis. Echocardiography, established, and bubble contrast study confirmed the diagnosis of an isolated cor-triatriatum dexter. The baby initially underwent an intervention by cardiac catheterization, which was unsuccessful in disrupting the membrane and re-direct the systemic venous flow to the right heart chambers. She subsequently had the cor-triatriatum dexter membrane resected via an uncomplicated open-heart surgery.

Keywords: Congenital heart disease, cor-triatriatum dexter, cyanosis, eustachian valve

INTRODUCTION

Cor-triatriatum dexter is an extremely rare congenital heart defect in which there is complete persistence of the right valve of embryonic sinus venosus that results in partitioning of the right atrium into a smooth and trabeculated portion. The smooth portion receives venous blood from inferior vena cava, superior vena cava, and coronary sinus while the trabeculated portion contains the right atrial appendage and the opening of tricuspid valve.

CASE REPORT

Our case is a 1-week-old child who was born after an uneventful pregnancy. Her Apgar score was 6 and 9 at one and 5 min, respectively. Her birth weight was 2.9 kg. Parents were first-degree relatives with no family history of congenital heart defects. She presented to the emergency department with intermittent episodes of circumoral cyanosis that was not related to feeding. The baby had no history of associated tachypnea or choking episodes. Her physical examination revealed central cyanosis with oxygen saturations between 85 and 88%, even with administration of 100% oxygen. The baby was otherwise well with no signs of respiratory distress. Her vital signs were within normal range for her age. Her cardiovascular examination showed a quite precordium, normal heart sounds, and no murmur. The rest of physical examination was unremarkable. Her investigations, including complete blood count, septic work-up, liver and renal function test, ammonia, and lactate, were all normal. Her electrocardiogram and chest X-rays were also unremarkable.

Echocardiography showed situs solitus, levocardia, atrio-ventricular and ventriculo-arterial concordance,

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and normal connection of pulmonary and systemic veins. There was a prominent and elongated membrane within the right atrium (RA) arising from the mouth of inferior vena cava (IVC) dividing the RA into two chambers. Both blood flow from IVC and superior vena cava (SVC) were directed by this membrane to the left atrium (LA) via a stretched patent foramen ovale (PFO) [Figure 1]. This membrane created a pouch that partially prolapsed across tricuspid valve (TV) during diastole. This pouch contained an opening through which some of the systemic venous blood was directed eccentrically across TV toward the right ventricle (RV) with no significant inflow gradient across TV [Figure 2]. The rest of the detailed echocardiography study did not reveal any other intra- or extra-cardiac abnormal findings and showed a good biventricular systolic function. Bubble contrast administered via a peripheral line connected to left forearm showed no left SVC and confirmed the diagnosis of cor-triatriatum dexter. The bubbles reached RA via the single right SVC and were directed by the cor-triatriatum dexter membrane toward LA via the PFO [Video 1].

Baby initially underwent cardiac catheterization performed by an experienced pediatric cardiac interventionist, but the procedure was unsuccessful to disrupt the cor-triatriatum dexter membrane and to re-direct the systemic venous flow toward TV. Subsequently, she underwent uncomplicated open-heart surgery at the age of 15 days during which the cor-triatriatum dexter membrane was resected. Her subsequent follow-up showed no residual membrane and there was laminar IVC and SVC flow across TV into a normal sized RV [Figure 3].

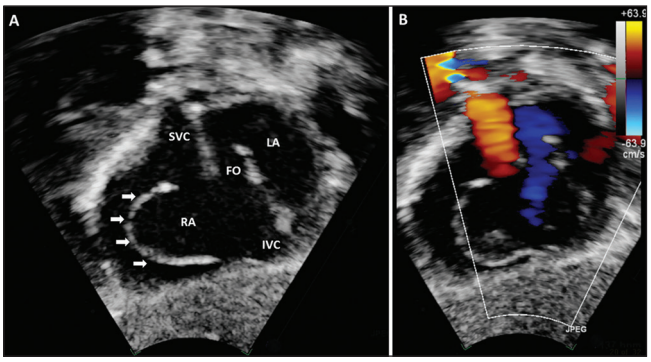

Figure 1: Two-dimensional (a) and two-dimensional color flow Doppler (b) echocardiography images from the sub-costal window. (a) The cor-triatriatum dexter membrane (arrows) that separates the right atrium into two parts. The inner part of right atrium receives the systemic venous blood from superior vena cava and inferior vena cava which is directed by the cor-triatriatum membrane to the left atrium via the foramen ovale. (b) The right to left shunt (blue color) across the foramen ovale. RA: Right atrium, SVC: Superior vena cava, IVC: Inferior vena cava, LA: Left atrium, FO: Foramen ovale

DISCUSSION

The remodeling of the RA during embryogenesis results from the incorporation of pectinated trabeculations of RA and the right horn of sinus venosus that consists of the right and left valvular folds in the sinoatrial orifice. The right valvular fold divides RA into two parts and directs IVC blood to LA through PFO.[1] The natural history of this valve is regression between 9 and 15 weeks of gestation leaving behind remnants such as the crista terminalis superiorly and the eustachian valve of IVC and the besian valve of the coronary sinus (CS) inferiorly. Complete persistence of the right sinus valve results in partitioning of RA into a smooth and trabeculated portion and constitutes the cor-triatriatum dexter. The smooth portion receives venous blood from IVC, SVC, and CS while the trabeculated portion contains the RA appendage and the opening of TV.[2]

Cor-triatriatum dexter can occur as an isolated cardiac anomaly as in our case or in association with other heart abnormalities.[3-6] In cor-triatriatum dexter, most of systemic venous blood is directed to LA through PFO, which explains the central cyanosis associated with this condition. Depending on the degree of flow obstruction at atrial or TV level, cor-triatriatum dexter might lead to right sided heart failure.[7]

Surgical resection of cor-triatriatum dexter is the treatment of choice with usually excellent short and long-term outcomes, although in stable and asymptomatic neonates with mild cyanosis this intervention may be deferred up to few months at other centers. Recently, percutaneous catheter disruption of the membrane has been reported and has been suggested as a preferred

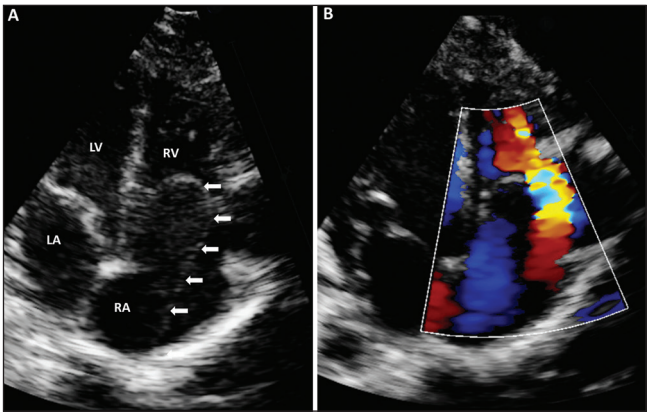

Figure 2: Two-dimensional (a) and two-dimensional color flow Doppler (b) echocardiography images in four chamber views from the apical window. (a) The cor-triatriatum dexter membrane (arrows) which is partially prolapsing across the tricuspid valve to the right ventricle during diastole. (b) Eccentric tricuspid valve inflow (red color) from the outer part of the right atrium to right ventricle. The blue color represents the systemic venous blood flow that is diverted back to right atrium by the cor-triatriatum dexter membrane. LA: Left atrium, LV: Left ventricle, RA: Right atrium, RV: Right ventricle

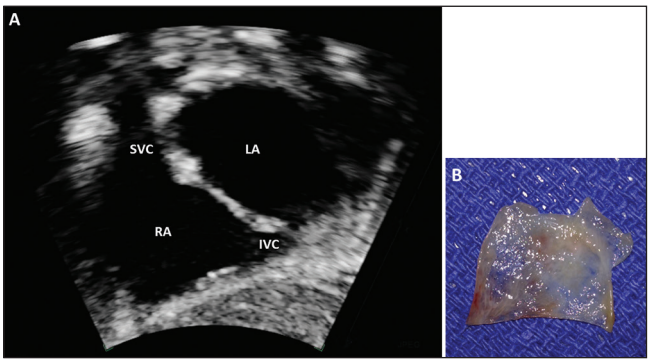

Figure 3: A demonstrates a two-dimensional echocardiography image from sub-costal window (a) after the surgical resection of the the cor-triatriatum dexter membrane and patch closure of the atrial septal defect. (b) A piece of the the cor-triatriatum dexter membrane. IVC: Inferior vena cava, LA: Left atrium, RA: Right atrium, and SVC: Superior vena cava

alternative to open-heart surgery though this procedure was not successful in our patient.[8]

We report this case to highlight cor-triatriatum dexter as a rare cause of central cyanosis during the neonatal period that can be missed since the episodes may occur intermittent. Diagnosis is established by a detailed and comprehensive echocardiography with a high index of suspicion. A bubble contrast echocardiography is not essential to establish the diagnosis of cor-triatriatum dexter, however, it may be helpful to exclude other rare causes of systemic venous abnormalities that can cause central cyanosis, such as persistent left SVC to left atrium, which can be missed by two-dimensional and color Doppler echocardiography alone.

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Conflicts of interest

There are no conflicts of interest.

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