Obstructing Eustachian valve in a newborn presenting with profound hypoxemia and atrial arrhythmias

Brian L. Wolfe, MD,[a] Elizabeth H. Stephens, MD, PhD,[b] Paul Tannous, MD, PhD,[a] Valerie Kalinowski, MD, MHA,[c] Amy S. Lay, MD,[a] Joseph M. Forbess, MD,[b] and Sandhya R. Ramlogan, MD,[a] Chicago, Ill

From the Divisions of [a]Cardiology and [b]Cardiovascular-Thoracic Surgery, Ann & Robert H. Lurie Children's Hospital of Chicago; and [c]Department of Pediatrics, Rush University Children's Hospital, Chicago, Ill.

Disclosures: The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

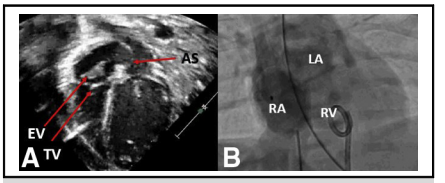
Received for publication March 18, 2020; revisions received March 18, 2020; accepted for publication April 2, 2020; available ahead of print April 11, 2020.

Address for reprints: Brian L. Wolfe, MD, 225 E Chicago Ave, Box 21, Chicago, IL 60611-2991 (E-mail: bwolfe@luriechildrens.org).

JTCVS Techniques 2020;3:265-6

2666-2507

https://doi.org/10.1016/j.xjtc.2020.04.008



Echo showing EV above TV (A) and angiogram showing contrast passing from RA to LA (B).

CENTRAL MESSAGE

Enlarged Eustachian valves may obstruct right ventricular inflow, causing hypoxemia secondary to right-to-left atrial level shunting.

See Commentary on page 267.

In fetal circulation, the Eustachian valve (EV) directs inferior vena cava blood (oxygenated blood from the placenta) across the patent foramen ovale (PFO). As the pulmonary vascular resistance drops after birth, this blood is no longer shunted to the left atrium and proceeds through the right heart to the lungs. However, in very rare instances, if the EV is large and nonfenestrated, it can cause postnatal obstruction to normal antegrade blood flow across the tricuspid valve. This leads to a large, persistent right-to-left shunt with subsequent systemic hypoxemia.

Our patient was a 13-day-old twin boy born at 35 weeks and 5 days' gestation. He developed hypoxemia in the delivery room and acidosis within 24 hours of life. He required progressive cardiopulmonary support and was ultimately placed on venoarterial extracorporeal membrane oxygenation (ECMO; via the right common carotid artery and right internal jugular vein) on the second day of life. In addition, he had bursts of supraventricular tachycardia (ectopic atrial tachycardia and atrial flutter) beginning at approximately 8 hours of life. The care team initially felt that both the hypoxia and arrhythmias were secondary to pulmonary hypertension. He was transferred to our institution on venoarterial ECMO and infusions of amiodarone and procainamide. After transfer, echocardiography demonstrated membranous tissue above the tricuspid valve (Figure 1, *A*), a small secundum atrial septal defect versus stretched PFO, and mildly hypoplastic right-sided structures (pulmonary valve and branch pulmonary arteries with z scores −2.3 to −2.8). The tricuspid valve annulus size was normal in the medial–lateral dimension (z score −1.2), but small in the anterior–posterior dimension (z score −3.4), and the right ventricle qualitatively lacked a normal trabecular portion. There was right-to-left shunting at the atrial level and apparent obstruction to right ventricular inflow.

A diagnostic cardiac catheterization was performed on day of life 15 while the patient was still on ECMO, but with dramatically reduced flows. This showed normal mean right atrial pressure (4 mm Hg), right ventricular systolic pressure (27 mm Hg—<1/2 systemic), and right ventricular end-diastolic pressure (6 mm Hg). Angiography showed a thin membrane above the tricuspid valve and preferential flow from the inferior vena cava to the left atrium (Figure 1, *B*).

The patient was taken to the operating room the next day, where he underwent resection of the EV, subtotal closure of the PFO, decannulation from ECMO, and reconstruction of the right common carotid artery. Antiarrhythmic medication was discontinued on postoperative day 2 and the patient experienced no further arrhythmias. The patient had an uneventful surgical recovery and was discharged home on postoperative day 23 on nasogastric feeds (for oropharyngeal aspiration), levothyroxine (for possible amiodarone-associated hypothyroidism), levetiracetam (due to concern for seizures while on ECMO), and aspirin (antiplatelet therapy following carotid artery reconstruction). At his most recent follow-up 6 weeks after surgery, the right atrial anatomy appeared normal, and his right-sided structures measured in the normal

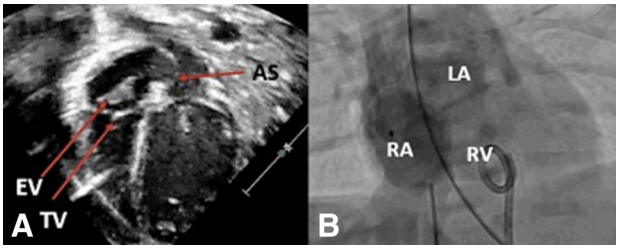


FIGURE 1. A, Apical 4-chamber echocardiogram image showing enlarged Eustachian valve (*EV*) just above the tricuspid valve (*TV*) with bowing of the atrial septum (*AS*) into the left atrium. B, Anteroposterior view during cardiac catheterization showing contrast preferentially entering the left atrium (*LA*) rather than the right ventricle (*RV*) with injection of contrast into the inferior vena cava.

range (tricuspid valve, pulmonary valve, and branch pulmonary arteries z scores –1.8 to +0.7).

In cor triatriatum dexter, there is a membrane originating from the valves of the embryonic sinus venosus separating the right atrium into a posteromedial chamber containing the caval veins and coronary sinus orifice and an anteromedial chamber containing the right ventricular inflow and the right atrial appendage. In this lesion, the amount of blood entering the right ventricle depends upon the number and size of fenestrations in the membrane as well as the size of any atrial septal defect. Although somewhat similar physiologically, our patient's lesion did not create separate chambers in the right atrium but did limit antegrade flow across the tricuspid valve. The intraoperative diagnosis was that of a large, nonfenestrated EV that almost reached to the plane of the atrial septum, significantly obstructing flow through the tricuspid valve. It measured approximately 1.5 cm × 1 cm (Figure 2).

Cor triatriatum dexter[1] or an enlarged EV[2,3] can both present with varying degrees of hypoxemia. We present a case of severe postnatal hypoxemia associated with refractory atrial arrhythmias due to a markedly enlarged and nonfenestrated EV. The vast majority of hypoxemic patients with

FIGURE 2. Image from operating room immediately postresection of Eustachian valve, with the tissue measuring approximately 1 cm × 1.5 cm.

otherwise structurally normal hearts have persistent fetal circulation/neonatal pulmonary hypertension. This case presents a rare mechanical cause for dramatic right-to-left shunting at the atrial level due to obstruction of tricuspid inflow. An obstructive EV or cor triatriatum dexter should remain in the differential diagnosis for patients such as this until definitively excluded with a focused anatomic evaluation. Fortunately, surgery appears to have been curative in this patient. The long-term prognosis, particularly the neurodevelopmental outcome, is unknown. Although the primary author's Institutional Review Board does not review/approve deidentified case reports such as this, our patient's mother and father have kindly provided verbal informed consent.

References

1. Mackman CA, Leidel JL, Woods RK, Samyn MM. A case series of cor triatriatum dexter: unique cause of neonatal cyanosis. *Pediatr Cardiol*. 2015;36:240-3.
2. Leon RL, Zaban NB, Schamberger MS, Ho CY, Mietzsch U. Cyanosis and stroke due to functional cor triatriatum dexter in a neonate. *Neonatology*. 2018;113:231-4.
3. Tamai K, Kageyama M, Takeuchi A, Nakamura M, Hirai K. Prominent Eustachian valve in a preterm infant causing cyanosis. *Pediatr Int*. 2018;60:311-3.