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CASE REPORT

BEGINNER

CLINICAL CASE

Obstructing Chiari Network Facilitating Blood Flow Across a Patent Foramen Ovale Causing Hypoxia

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ABSTRACT

A 36-year-old man with progressive dyspnea and hypoxia was found to have a large, partially fenestrated Chiari network accelerating blood flow through a patent foramen ovale with preservation of an embryonic right-to-left atrial flow pattern. He underwent successful percutaneous patent foramen ovale closure with resolution of his exertional symptoms. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:1025–8) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 36-year-old man incarcerated at the Adult Correctional Institutions site in Rhode Island, with a history of metabolic syndrome, presented with approximately 2 years of chronic dyspnea on exertion, associated nonproductive cough, and intermittent substernal chest discomfort. He also related episodes of paroxysmal nocturnal dyspnea, coughing and choking arousals several times weekly while recumbent, worsening daytime somnolence, and occasional headaches. He was known at the Adult Correctional Institutions to snore heavily. He denied sore throat, sinus pressure, orthopnea, uncontrolled heartburn or reflux, abdominal pain, peripheral edema, and weight gain. He recalled being diagnosed with a "hole in his heart" during childhood but denied any deleterious clinical sequelae related to this. He denied a history of stroke. He had normal vital signs at rest with an unremarkable physical examination. However, he became hypoxemic to 88% with minimal ambulation in 4-point shackles, requiring several minutes to improve back to baseline of 94% to 96%.

LEARNING OBJECTIVES

- To consider right-to-left intracardiac shunting in cases of unexplained hypoxemia.
- To understand that a physiologically important right-to-left intracardiac shunt can be caused by a PFO combined with a large residual Chiari network directing venous return toward a PFO and obstructing the normal flow pattern toward the right ventricle.

MEDICAL HISTORY

The patient had a history of hypertension, insulin-dependent diabetes mellitus, dyslipidemia, unspecified mood disorder with psychotic features, sickle-cell trait, gastroesophageal reflux disease, and

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

PFO = patent foramen ovale

hepatic steatosis. There was no family history of premature coronary artery disease, asthma, chronic obstructive pulmonary disease, or cirrhosis.

DIFFERENTIAL DIAGNOSIS

Given the patient's worsening dyspnea and limited work-up to date, the differential diagnosis for his unexplained hypoxemia remained broad. This included adult congenital heart diseases causing right-to-left shunting such as transposition of the great arteries, tricuspid disease with an atrial septal defect or ventricular septal defect including Ebstein's anomaly, tetralogy of Fallot, Taussig-Bing anomaly, truncus arteriosus, total anomalous pulmonary venous return with atrial septal defect, and Eisenmenger syndrome associated with atrial septal defect, ventricular septal defect, and patent ductus arteriosus. Pulmonary causes for hypoxia included pulmonary arteriovenous fistula and hepatopulmonary syndrome. Pulmonary causes of ventilation-perfusion mismatch such as pulmonary hypertension, interstitial lung disease, and chronic pulmonary emboli were also considered. Finally, the differential diagnosis included platypnea-orthodeoxia syndrome.

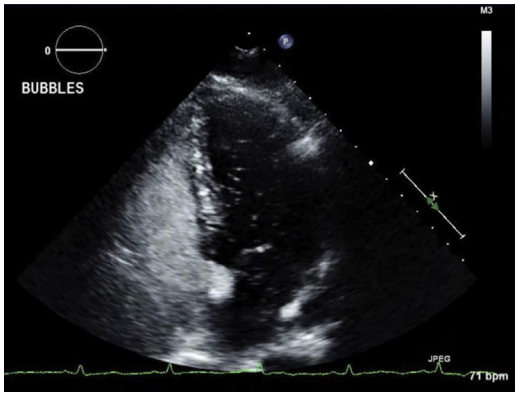
INVESTIGATIONS

Laboratory data were notable for abnormal resting arterial blood gas (pH = 7.37, pCO_2 = 45 mm Hg, pO_2 = 50 mm Hg) and hemoglobin of 14 g/dl. His electrolyte, serial troponin, and B-type natriuretic peptide levels were normal. Initial imaging demonstrated normal results on chest radiography and myocardial perfusion imaging. Transthoracic echocardiography was unremarkable.

After the index hospitalization, the patient was empirically treated with bronchodilators and discharged to the Adult Correctional Institutions with close pulmonology follow-up. His symptoms persisted and he was rehospitalized. At this time, pulmonary function tests demonstrated a reduced ratio of forced expiratory volume in 1 s to forced vital capacity and diffusion capacity of the lungs for carbon monoxide suggestive of restrictive lung disease, but high-resolution chest computed tomography ruled out intraparenchymal pulmonary fibrosis. There was now concern for a chronic vascular and/or intracardiac shunting process given his ongoing intermittent hypoxemia.

In the respiratory clinic, the patient reported no improvement with his inhalers, and hypoxemia was documented on a 6-min walk test. At this time, repeat transthoracic echocardiography with a bubble study (**Figure 1**, Video 1) noted a patent foramen ovale (PFO) with a large right-to-left shunt and an atrial septal aneurysm. Pulmonary pressures were normal. As such, he underwent transesophageal echocardiography, which was notable for an atrial septal aneurysm with a large PFO (**Figure 2**, Video 2) with left-to-right flow by color Doppler and right-to-left flow by saline contrast. A large fenestrated residual Chiari complex was also observed (**Figure 3**, Video 3), with flow acceleration across this membrane suggesting some degree of obstruction.

FIGURE 1 Large Patent Foramen Ovale on Bubble Study



Bubble study with agitated saline on transthoracic echocardiography demonstrating the presence of a patent foramen ovale.

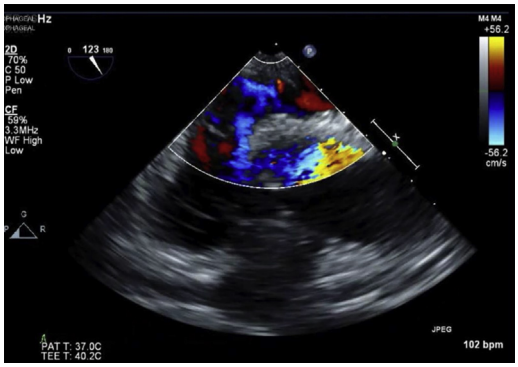
MANAGEMENT

The patient was scheduled for PFO closure to definitively address his persistent hypoxemia. Intraoperative echocardiography showed a PFO with an atrial septal aneurysm with bidirectional shunting at baseline. His hemodynamic pressures were normal. He underwent successful transfemoral percutaneous PFO closure with no post-procedural shunting noted by color Doppler flow. His oxygen saturation was 93% to 95% on 2 l supplemental oxygen pre-procedure and improved to 100% on 2 l oxygen post-procedure. He was discharged on aspirin 81 mg/day and clopidogrel 75 mg/day.

DISCUSSION

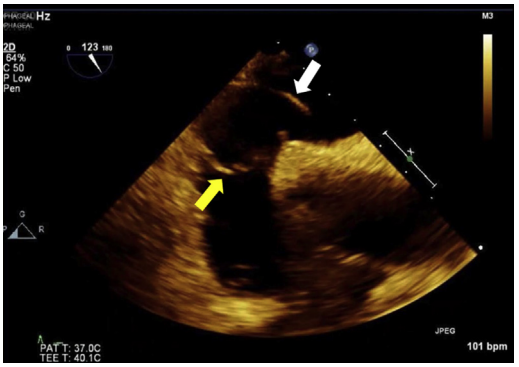
A Chiari network is a fenestrated network of fibers in the right atrium that are in connection with the eustachian and Thebesian valves at the opening of

FIGURE 2 Transesophageal Echocardiography With Color Doppler at Patent Foramen Ovale



Transesophageal echocardiography with Doppler color flow across the interatrial septum at the patent foramen ovale.

FIGURE 3 Transesophageal Echocardiography With Chiari Network



White arrow points to the Chiari network. **Yellow arrow** points to the interatrial septum.

the inferior vena cava and the coronary sinus that results from an incomplete resorption of the remnants of the right valve of the sinus venosus during embryonic development (1). This network usually has no clinical significance, but there have been reports of its role in forming thrombi, atrial septal aneurysms, facilitating paradoxical emboli, arrhythmias, and infective endocarditis. The network also contributes to difficult placement of devices such as catheters, guidewires, and pacemakers and acts as a protective filter against pulmonary emboli (2,3).

Of note, the Chiari network is associated with the persistence of a PFO, as the fibers may direct blood flow from the inferior vena cava to the interatrial septum, causing a right-to-left shunt (1). Generally, flow across an interatrial septal defect is left to right, but transient right to left shunting can occur, particularly with isometric strain (4) such as with a Valsalva maneuver or coughing. Right-to-left shunting is also associated with high pulmonary pressures as well as other anatomic changes such as thoracic aortic aneurysms, thoracic trauma with tricuspid regurgitation, and mediastinal shifts after thoracic surgery (5). Right-to-left shunting, although not as common as left-to-right shunting, can lead to profound hypoxemia.

In an asymptomatic patient, a Chiari network requires no intervention, but surgical resection can be considered if there is persistent hypoxemia, cyanosis, or obstruction (6). Similarly, an incidentally discovered PFO requires no follow-up or treatment unless it has high-risk features. Cryptogenic stroke is associated with a high-risk PFO score (≥2), which includes a Chiari network (7). However, PFO closure has been found to be beneficial in a variety of cases, such as reducing recurrent stroke or transient ischemic attacks compared with medical therapy (8), alleviating symptoms in patients with platypnea-orthodeoxia (9), and can potentially improve quality of life in patients with chronic obstructive pulmonary disease with hypoxemia (10).

In our patient with unexplained hypoxemia, who was found to have a large residual Chiari network in the setting of a large PFO, it was believed that he was having intermittent hypoxemia secondary to the structural cardiac defects. As such, he underwent PFO closure.

Right heart catheterization was not performed to quantify the right-to-left shunt, as it would not have changed clinical decision making. There was an anatomic reason for significant shunting, directing us to PFO closure. Furthermore, calculation of the shunt (Q_p/Q_s) in this case would be prone to errors. Systemic arterial flow (Q_s) would be calculated using Fick's principle and pulmonary flow (Q_p) using thermodilution from the catheterization. However, much of the temperature bolus from the saline could be directed by the Chiari network through the PFO and reduce the peak of the time/temperature curve seen by the thermistor in the pulmonary artery, resulting in a falsely elevated circulation of pulmonary flow.

FOLLOW-UP

The patient's PFO closure was successful, without post-procedural complications. He had resolution of his exertional symptoms at a 1-month post-procedure follow-up visit.

CONCLUSIONS

This case illustrates an unusual anatomic variation of two commonly benign findings on echocardiography, a Chiari network and PFO. These findings resulted in clinically significant and late presenting cyanotic heart disease with right-to-left shunting and should be considered in the spectrum of late presenting cyanotic heart disease in adults. Marked clinical improvement resulted from percutaneous PFO closure.

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KEY WORDS Chiari network, congenital heart defect, dyspnea, hypoxemia

APPENDIX For supplemental videos, please see the online version of this paper.