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Brittany Hartman

Albert Einstein Medical Center, brittany.hartman@jefferson.edu

Alex Dwyer

Albert Einstein Medical Center

Robert Dejoy

Albert Einstein Medical Center

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Visual Case Discussion

There's a hole in my heart: A case of an unrepaired VSD

Brittany Hartman^{a,*}, Alex Dwyer^a, Robert DeJoy^b^a Department of Emergency Medicine, Albert Einstein Medical Center, Philadelphia, PA, United States of America^b Department of Internal Medicine, Albert Einstein Medical Center, Philadelphia, PA, United States of America

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1. Introduction

Ventricular septal defects (VSD) account for almost 50% of all cases of congenital heart disease, and are the second most common congenital heart defect seen in adults.¹ Ventricular septal defects range in their location and size, and are commonly associated with other congenital heart defects. As much as 80% of all VSDs are tiny to small defects that are rarely associated with hemodynamic compromise or pulmonary hypertension² and as many as 90% of VSDs close spontaneously. Perimembranous VSDs are the most common type of VSD,³ and only rarely lead to hemodynamic compromise. Here, we present a case of an unrepaired perimembranous VSD in a 40-year-old male with pulmonary hypertension, left ventricular hypertrophy, and severely reduced ejection fraction leading to heart failure.

2. Case report

A 40-year-old male with a known history of unrepaired VSD presented to an urban emergency department for evaluation of worsening dyspnea, lower extremity edema, and fatigue. The patient had limited access to medical care, and had not seen a physician in more than 2 years at the time of his presentation. In the emergency department, the patient was found to have a loud, blowing, holosystolic murmur with 3+ pitting edema to bilateral lower extremities. Laboratory analysis was notable for a BNP >600, troponin of 0.03, and elevated creatinine level of 1.48. His-EKG showed prolonged QTC with significant biatrial enlargement (Fig. 1). Point-of-care ultrasound (POCUS) was performed at this time, which showed an obvious, moderately sized VSD with notable left

ventricular hypokinesis (Fig. 2). The patient was admitted to the telemetry service for management of the patient's heart failure, and he underwent a formal echocardiogram, showing severe eccentric left ventricular hypertrophy, perimembranous VSD with left to right shunt, and severely reduced left ventricular ejection fraction of 25%. He was also noted to have severely elevated pulmonary arterial pressures representing pulmonary hypertension. Following diuresis and optimal medical management of patient's heart failure exacerbation, the patient presented to the outpatient cardiothoracic surgery service for evaluation of repair of his clinically significant VSD leading to heart failure. The patient was pending repair at the time of this case report.

3. Discussion

The vast majority of VSDs close spontaneously without significant hemodynamic effects or clinical implications (3). VSDs that do not close spontaneously and are not repaired prior to adulthood are rarely associated with heart failure (2). In this patient, his VSD failed to close spontaneously and had not been surgically repaired, which led to clinically significant early onset heart failure with severely reduced ejection fraction and associated pulmonary hypertension. The emergency department team can serve as an early identifier of hemodynamically significant VSD leading to earlier referral and treatment. There are many screening tools available to the emergency department provider including history and physical, EKG, chest x-ray and ultimately POCUS (3). Of these screening tools POCUS is the only modality capable of visualizing the defect while the others serve to identify left heart failure (Fig. 2).

Abbreviations: VSD, Ventricular Septal Defect; CHF, Congestive heart failure.

* Corresponding author at: 5501 Old York Rd, Philadelphia, PA, 19144, United States of America.

E-mail address: brittany.hartman@jefferson.edu (B. Hartman).

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4. Visual case discussion

POCUS can serve as an early screening tool for patients with dyspnea, and furthermore, ultrasound is the only imaging modality that can directly visualize a VSD. On history and physical symptoms of shortness of breath, orthopnea and paroxysmal nocturnal dyspnea may point to left heart failure in addition to a pan-systolic murmur over the left lower sternal border and rales on physical exam. EKG can also show evidence of left heart dysfunction exhibited by left ventricular hypertrophy or evidence of right heart strain indicating pulmonary arterial hypertension, as well as prolonged QTc (Fig. 1). Chest x-ray can demonstrate an enlarged cardiac silhouette which could represent left ventricular hypertrophy. However, POCUS may be the only tool the emergency medicine provider has to directly visualize the VSD at bedside. Given the significant size of the patient’s VSD and the ease with which it was seen on POCUS, the patient may have benefited from earlier recognition of the lesion had he had an early POCUS or echocardiogram. Earlier recognition can aid in referral and definitive management however in the emergency department these patients should be emergently treated as any other patient presenting with evidence of heart failure. Nevertheless, earlier recognition could have aided in management and ultimately prevented development of the clinical sequelae associated with a persistent unrepaired VSD.

5. Conclusion

POCUS is an important adjunct that can be used in the emergency department for evaluation of cardiac function. In this case, the patient’s VSD was large enough to be easily visible on an apical 4-chamber POCUS view of the heart. This patient had clinically significant heart failure associated with his VSD which could have been addressed sooner had POCUS been utilized during previous visits.

6. Mandatory questions and answers

1. Which valvular abnormalities are adult patients with an unrepaired VSD most at risk for?



Fig. 2. A four-chamber view of the heart demonstrating a moderately sized VSD with left ventricular hypertrophy.

- A Aortic regurgitation
- B Pulmonic stenosis
- C Pulmonic regurgitation
- D Mitral valve prolapse

Answer: A.

Discussion: The left or right aortic valve cusp can be less effective secondary to hypoplasia of the septum. Early surgical repair is recommended in patients with aortic regurgitation secondary to a VSD.

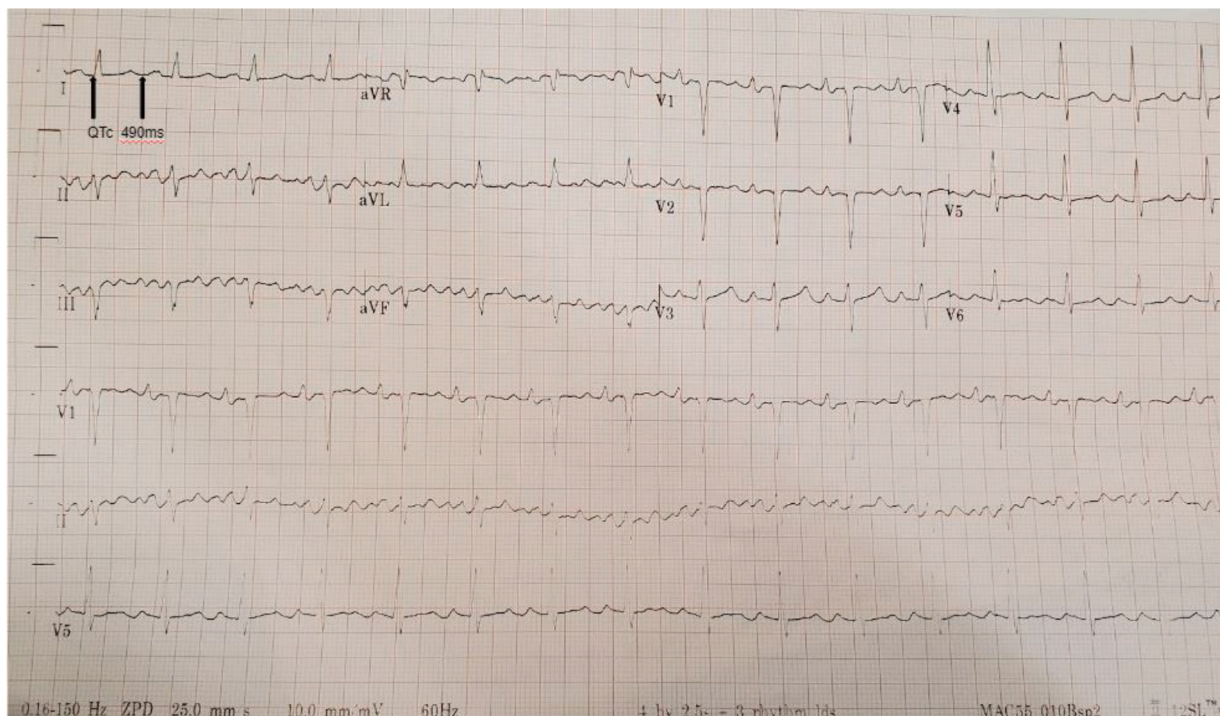


Fig. 1. EKG demonstrating prolonged QTc with significant biatrial enlargement.

Ammash N. et al. Management and Prognosis of Ventricular Septal Defects in Adults. UptoDate. Retrieved June 3, 2022, from <https://www.uptodate.com/contents/management-and-prognosis-of-ventricular-septaldefect-in-adults?search=ventricular-septal-defect-in-adult>

2. Which is NOT an indication for VSD closure in an adult?

- A Adults with VSD and worsening aortic regurgitation
- B Adults with history of infective endocarditis
- C Adults with nonrestrictive VSD and evidence of Eisenmenger syndrome
- D Adults with VSD without evidence of pulmonary hypertension

Answer: C

Discussion: Patients who develop Eisenmenger syndrome secondary to a patent VSD benefit from dual heart and lung transplantation as treatment however fare worse after isolated VSD treatment alone. The VSD itself is thought to be protective and prevents further increase in pulmonary vascular resistance through shunting. Closure of VSDs in Eisenmenger syndrome has been shown to lead to right ventricular failure and pulmonary hypertensive crisis therefore optimal medical management is recommended over surgical repair in these patients.

CRediT authorship contribution statement

Brittany Hartman: Conceptualization, Writing – original draft, Supervision. **Alex Dwyer:** Writing – review & editing, Visualization. **Robert DeJoy:** Writing – review & editing, Investigation.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at [doi:10.1016/j.visj.2022.101581](https://doi.org/10.1016/j.visj.2022.101581).

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