CLINICAL VIGNETTE

A Rare Case of Atrial Septal Defect Presenting as Postpartum Pulmonary Hypertension

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Introduction

Atrial septal defect (ASD) is one of the most common congenital heart diseases in adults.[1] In many instances, it is diagnosed during or shortly after pregnancy. The hemodynamic changes which occur during pregnancy can exacerbate underlying cardiac conditions, and in turn unmask a pre-existing condition.[2] We present a case of a young, post-partum female who presented with symptoms of heart failure due to a large ASD.

Case Presentation

A 23-year-old female with no known past medical history presented to the emergency department with atypical chest pain and shortness of breath. She reported that she had delivered her first baby 2 weeks prior at an outside hospital and had been experiencing symptoms since delivery. Symptoms were noted after delivery at the outside hospital and she was discharged on furosemide and with supplemental oxygen. In the emergency department the patient was hypoxic at 91% on room air. Electrocardiogram showed right ventricle (RV) hypertrophy, incomplete right bundle branch block, and a first-degree AV block. Her EKG also had notching of the QRS in the inferior leads. Troponin was negative, but CT pulmonary angiography showed markedly dilated pulmonary arteries (PA) as well as significant right atrial (RA) and RV dilation consistent with right heart strain. A transthoracic echocardiogram displayed a large secundum atrial septal defect measuring 5.3 cm in diameter with a bidirectional atrial shunt. She was also noted to have a severely dilated right ventricle, and moderate pulmonary and tricuspid valve regurgitation with pulmonary artery pressure of 67mmHg. Right heart catheterization noted severely elevated pulmonary arterial pressure (70/24 mmHg), with normal RA pressure and left sided hemodynamics. The pulmonary flow to systemic flow ratio (Qp/Qs) was 1.35 using hemodynamic measurements. Pulmonary to systemic systolic blood pressure ratio (PASP:SBP) was 0.54 (70/129). Cardiothoracic surgery was consulted, and patient underwent ASD closure. Her symptoms improved after the surgery and she was discharged without diuretics and she remained asymptomatic at her follow-up clinic appointment.

Discussion

ASD is one of the most common congenital diseases in adults. It is important to diagnose patients early, as risk of morbidity and mortality increase if left untreated. The primary test to diagnose an ASD is a transthoracic echocardiogram (TTE). An EKG for an ostium secundum ASD may show “crochetage sign” (notching of R in the QRS complex of inferior leads), as seen in this case. ASD results in shunting of blood between systemic and pulmonary circulation. Initially shunting occurs from the left to right side of the heart, due to higher chamber pressures on the left side of the heart**,** with the degree of shunting directly correlated to the size of the ASD. This shunting leads to excessive blood flow in the RV and PA, and in turn causes RV and RA dilation.[3] Over time, the dilation may result in RV failure, pulmonary hypertension, arrhythmias, and paradoxical embolism. The risk of pulmonary vascular disease increases with age and female gender.[1] Symptoms of ASD are non-specific at time of presentation, and include chest pain, fatigue, and exertional dyspnea.

Pregnancy causes specific physiologic changes that affect hemodynamics and can unmask an ASD. Firstly, hormonal changes early in pregnancy decrease blood pressure by 5-10 mmHg, leading to an increase in cardiac output. Secondly, plasma volume can increase by up to 50% through the second trimester, also increasing cardiac output.[4] These physiologic changes can exacerbate RV overload, especially in women with RV dilation and enlargement from untreated ASDs, leading to the characteristic symptoms of heart failure described above.

The postpartum period is also associated with significant hemodynamic changes. Within 10 minutes following vaginal delivery, cardiac output and stroke volume can increase by 60 to 70 percent. These changes most likely result from increased cardiac preload from auto transfusion of placental blood to the intravascular space. In addition, the decompression of inferior vena cava post-delivery results in increased preload. These changes can result in unmasking of a previously asymptomatic ASD.[5]

AHA guidelines recommend closure of secundum ASD causing impaired functional capacity, RA or RV enlargement, and net left to right shunt sufficiently large to cause physiological sequelae (Qp/Qs >1.5). Closure is recommended as long as systolic pulmonary artery pressure is less than 50% of systolic systemic pressure and pulmonary vascular resistance is less than 33% of the systemic vascular resistance.[6] In more advanced disease with pulmonary hypertension, the ASD serves as a pop off valve. In cases where the pulmonary to systemic

pressure ratio is more than 50%, closure of the ASD could lead to right sided pressure overload and is not recommended.

Treating an ASD in pregnancy depends on when it is diagnosed. Most women newly diagnosed with ASD during pregnancy can have closure deferred until six-months after delivery. In cases where an ASD is known in advance, closure is recommended before pregnancy. Further, if the known ASD has resulted in severe pulmonary hypertension, pregnancy is contraindicated as it can result in high morbidity and mortality for the mother and fetus. In general, pregnancy is well tolerated in most women with uncomplicated ASDs.[7]

Our case was different because the patient did not meet the strict criteria for surgical closure. The Qp/Qs ratio was less than 1.5 and her PASP/SBP was slightly greater than 50%. However, she was clearly symptomatic from the ASD and she also had some RV dilation. There was concern that deferring surgical correction of the ASD would result in a net right to left shunt and subsequent Eisenmenger syndrome, a feared complication of an untreated ASD. After a discussion with the patient, the decision was made to proceed with ASD closure with the hope of reducing symptoms and preventing further RV dysfunction. The patient's symptoms resolved after closure.

Conclusion

Atrial septal defect is one of the most common congenital heart disease in adults and can go undiagnosed into adulthood. It is important to diagnose patients early to prevent development of pulmonary hypertension and right ventricular failure. The hemodynamic changes during pregnancy and post-partum can unmask underlying, undiagnosed cardiac conditions, as in this case. Once diagnosed, the decision to close ASD is complicated, dependent on multiple factors, and requires adequate risk/benefit analysis, patient communication, and a multi-disciplinary approach.

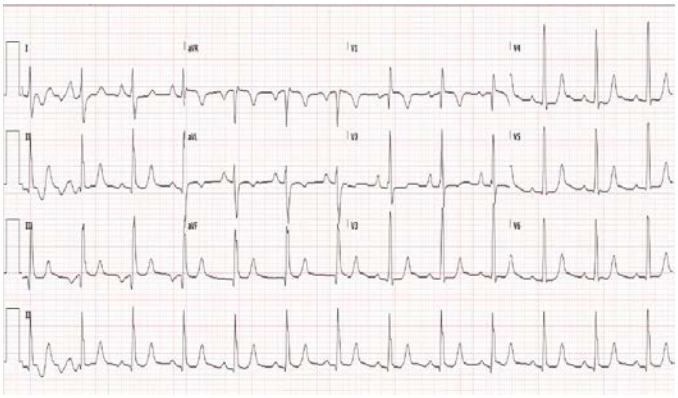

Figure 1. EKG showing crochetage sign with R notching in the inferior leads.

REFERENCES

1. **Geva T, Martins JD, Wald RM**. Atrial septal defects. *Lancet*. 2014 May 31;383(9932):1921-32. doi: 10.1016/S0140-6736(13)62145-5. Epub 2014 Apr 8. PMID: 24725467.
2. **Ouzounian JG, Elkayam U**. Physiologic changes during normal pregnancy and delivery. *Cardiol Clin*. 2012 Aug;30(3):317-29. doi: 10.1016/j.ccl.2012.05.004. Epub 2012 Jun 20. PMID: 22813360.
3. **Beitzke D, Wolf F, Edelhauser G, Lammer J, Loewe C**. Right heart dilatation in adult congenital heart disease: imaging appearance on cardiac magnetic resonance. *Br J Radiol*. 2011 Feb;84(998):188-93. doi: 10.1259/bjr/13711325. Epub 2010 Dec 15. PMID: 21159804; PMCID: PMC3473844.
4. **Soma-Pillay P, Nelson-Piercy C, Tolppanen H, Mebazaa A**. Physiological changes in pregnancy. *Cardiovasc J Afr*. 2016 Mar-Apr;27(2):89-94. doi: 10.5830/CVJA-2016-021. PMID: 27213856; PMCID: PMC4928162.
5. **Ueland K, Hansen JM**. Maternal cardiovascular dynamics. II. Posture and uterine contractions. *Am J Obstet Gynecol*. 1969 Jan 1;103(1):1-7. doi: 10.1016/s0002-9378(16)34333-2. PMID: 5761774.
6. **Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, Crumb SR, Dearani JA, Fuller S, Gurvitz M, Khairy P, Landzberg MJ, Saidi A, Valente AM, Van Hare GF**. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Circulation*. 2019 Apr 2;139(14):e698-e800. doi: 10.1161/CIR.0000000000000603. Erratum in: Circulation. 2019 Apr 2;139(14):e833-e834. PMID: 30586767.
7. **Bredy C, Mongeon FP, Leduc L, Dore A, Khairy P**. Pregnancy in adults with repaired/unrepaired atrial septal defect. *J Thorac Dis*. 2018 Sep;10(Suppl 24):S2945-S2952. doi: 10.21037/jtd.2017.10.130. PMID: 30305955; PMCID: PMC6174140.