Diastolic Dysfunction Manifesting as Acute Plastic Bronchitis Following Warden Procedure

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PII: S2666-2507(24)00147-0
DOI: https://doi.org/10.1016/j.xjtc.2024.03.018
Reference: XJTC 1659

To appear in: JTCVS Techniques

Received Date: 21 January 2024
Revised Date: 19 February 2024
Accepted Date: 13 March 2024

Please cite this article as: Pasternack DM, Martinez MJ, McKinstry J, Singh R, Saharan S, Muise ED, Mosca R, Kumar TKS, Diastolic Dysfunction Manifesting as Acute Plastic Bronchitis Following Warden Procedure, JTCVS Techniques (2024), doi: https://doi.org/10.1016/j.xjtc.2024.03.018.

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Diastolic Dysfunction Manifesting as Acute Plastic Bronchitis Following Warden Procedure

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Author Disclosure: Drs. Pasternack, Martinez, McKinstry, Singh, Saharan, Muise, Mosca, and Kumar have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

Funding Statement: This manuscript received no external funding.

Consent: The subject’s legal guardian provided informed consent for the publication of this report.

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Word Count: 717
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Central Image Legend: Bronchial casts suggestive of plastic bronchitis extracted during flexible bronchoscopy to evaluate lung opacification.
Central Message (200 character limit)

Atrial septal defects (ASD) can mask the presence of diastolic dysfunction (DD). DD should be considered in any pre-tricuspid shunt when the left ventricle is disproportionately small.
Introduction

Left ventricular (LV) diastolic dysfunction (DD) is well-recognized in adult patients with atrial septal defects (ASDs).\textsuperscript{1} Chronic underfilling of the LV combined with altered geometry caused by right ventricular (RV) volume overloading, leads to the LV DD. This can manifest as acute pulmonary edema following closure of the ASD. However, this phenomenon is rarely witnessed in children.\textsuperscript{2} We report the case of a child who developed plastic bronchitis due to unrecognized DD following a Warden Procedure for a sinus venosus ASD (svASD) with partially anomalous pulmonary venous connection (PAPVC).

Case Report

The subject’s legal guardian provided informed consent for the publication of this report; IRB approval was not required. A 10-year-old male with sickle cell trait, and a recent immigrant from Haiti, presented for the evaluation of a murmur. He was found to have a large svASD with the right upper pulmonary vein draining into the superior vena cava (SVC). His LV end-diastolic volume Z-score was -3.23. He was largely asymptomatic with only mild exercise intolerance.

Surgical repair of the svASD and PAPVC using a single-patch technique was attempted but was converted to a Warden procedure due to concerns for SVC narrowing. Post-procedure echocardiogram demonstrated excellent repair with unobstructed systemic and pulmonary venous return. He was extubated in the operating room and transferred to the intensive care unit for further management.

Later that day he developed respiratory distress requiring reintubation. Chest radiograph demonstrated bilateral pulmonary infiltrates. A second extubation attempt failed due to recurrent pulmonary edema despite medical optimization. Near-complete opacification of the left lung was noted (supplemental figure 1). Flexible bronchoscopy demonstrated a significant cast burden throughout the bronchial tree (figure 1) consistent with plastic bronchitis. Serial echocardiograms continued to demonstrate normal systolic function.

Cardiac catheterization demonstrated no baffle leak and a trivial SVC gradient, although end-diastolic pressure (EDP) was elevated in both right and left ventricles (16 and 25 mm Hg respectively; figure 2). Following a detailed discussion, transseptal puncture of the fossa ovalis with balloon atrial septoplasty was performed, decreasing left ventricular EDP by 4 mmHg. Diuretic and lusitropic therapy with milrinone were also initiated.

His condition improved, and he was successfully extubated in the next few days. He was slowly weaned off all respiratory and vasoactive support and was discharged home after a three-week hospitalization. Repeat catheterization four months later demonstrated interval improved biventricular filling pressures under baseline conditions (figure 2). However, a fluid challenge demonstrated persistent DD. Now one year post-operatively, he has remained clinically well without medical therapy and is able to participate in recreational sports.
Discussion

DD can be masked by the presence of an ASD. The left atrial hypertension that results from poor left ventricular compliance is offset by the left-to-right shunt at the atrial level. Thus, an ASD will mitigate the onset of DD symptoms. The presence of PAPVC further reduces the amount of venous return to the left atrium. Herein, we describe the case of a patient whose DD was unmasked by ASD closure and PAPVC repair, resulting in pulmonary edema and respiratory failure.

In children, DD is rare, and diagnosis can be easily missed by conventional noninvasive testing.\(^2\) Left atrial enlargement, the most sensitive echocardiographic finding in DD, is not seen with an ASD.\(^3\) Direct measurement EDP via catheterization remains the gold standard of diagnosis.\(^3\) The signs of DD may be subtle early in the course of the disease, and may be missed by invasive and noninvasive modalities.\(^4\) Such investigation is essential, as pre-operative evidence of DD has been associated with post-operative heart failure.\(^1\)

Our case highlights the importance of assessing for DD prior to ASD closure. Although the LV size is generally small in the setting of ASD, one must suspect the presence of coexisting DD when the LV size is disproportionately smaller. If pre-operative echocardiographic evidence of LV hypoplasia or DD is present, further evaluation including a cardiac catheterization or intraoperative left atrial pressure measurement should be pursued.\(^1\) Other causes of DD include chronic microinfarctions from sickle cell anemia, although this happens later in life and has not been reported with sickle cell trait.\(^5\)

Conclusion

A high index of suspicion for DD should be entertained in patients with any pre-tricuspid shunt when the left ventricular size is disproportionately small. Repair of the defect in such settings can lead to hemodynamic and respiratory compromise, as occurred in our patient.


Tables

[None]

Figure Legends

Figure 1. Bronchial casts suggestive of plastic bronchitis extracted during flexible bronchoscopy.

Figure 2. Baseline left ventricular pressure tracing from the initial post-operative (A, B) and follow-up (C, D) cardiac catheterizations.

Supplemental Figure 1. Chest radiograph demonstrating complete opacification of left lung, observed following unsuccessful attempts at extubation in the first few post-operative days.