

Diastolic dysfunction manifesting as acute plastic bronchitis after Warden procedure



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Bronchial casts extracted during flexible bronchoscopy to evaluate lung opacification.

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

CASE REPORT

The subject’s legal guardian provided informed consent for the publication of this report; institutional review board approval was not required. A 10-year-old boy 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 because of concerns for SVC narrowing. Postprocedure 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. Radiograph of the chest demonstrated

CENTRAL MESSAGE

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

bilateral pulmonary infiltrates. A second extubation attempt failed because of recurrent pulmonary edema despite medical optimization. Near-complete opacification of the left lung was noted (Figure E1). 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). After a detailed discussion, transeptal puncture of the fossa ovalis with balloon atrial septoplasty was performed, decreasing left ventricular EDP by 4 mm Hg. 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 3-week hospitalization. Repeat catheterization 4 months later demonstrated interval improved biventricular filling pressures under baseline conditions (Figure 2). However, a fluid



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

challenge demonstrated persistent DD. Now 1 year postoperatively, 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 LV 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 preoperative evidence of DD has been associated with postoperative heart failure.¹

Our case highlights the importance of assessing for DD before 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 preoperative 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

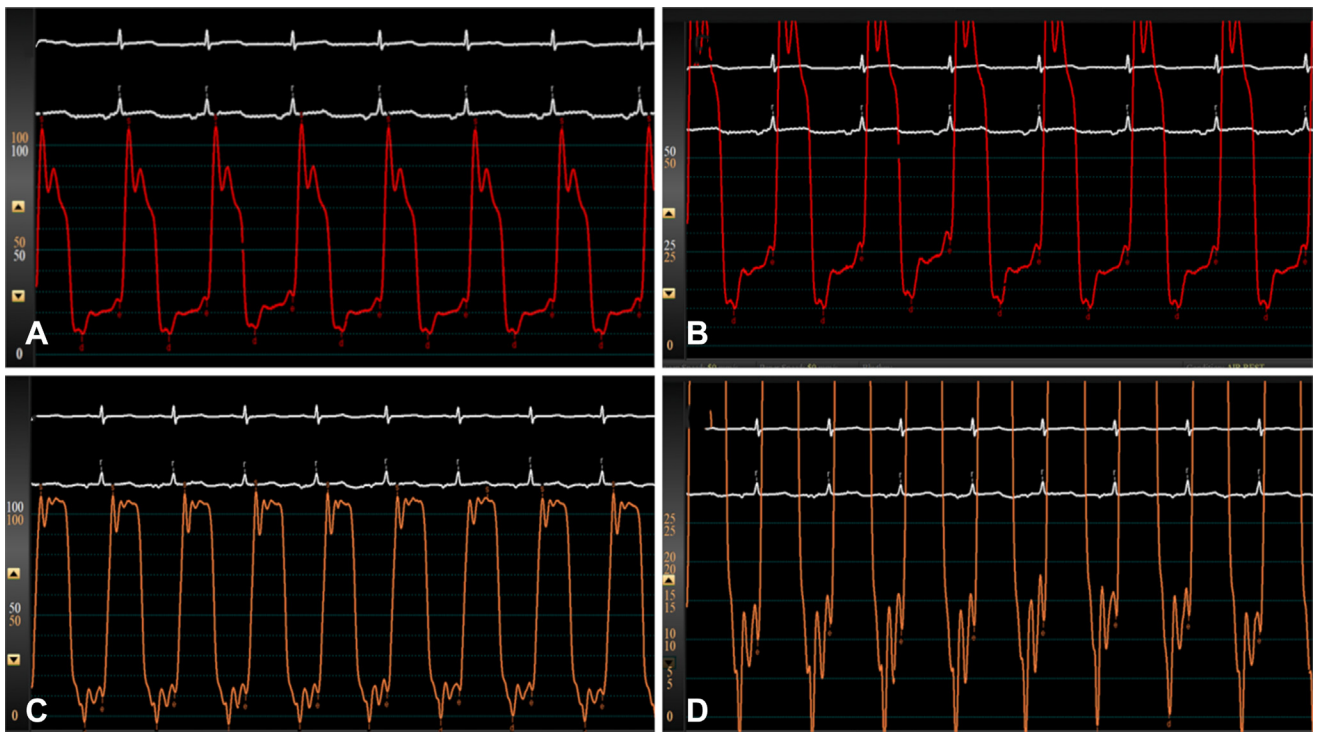


FIGURE 2. Baseline left ventricular pressure tracing from the initial postoperative (A, B) and follow-up (C, D) cardiac catheterizations.

CONCLUSIONS

A high index of suspicion for DD should be entertained in patients with any pretricuspid 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.

Conflict of Interest Statement

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.

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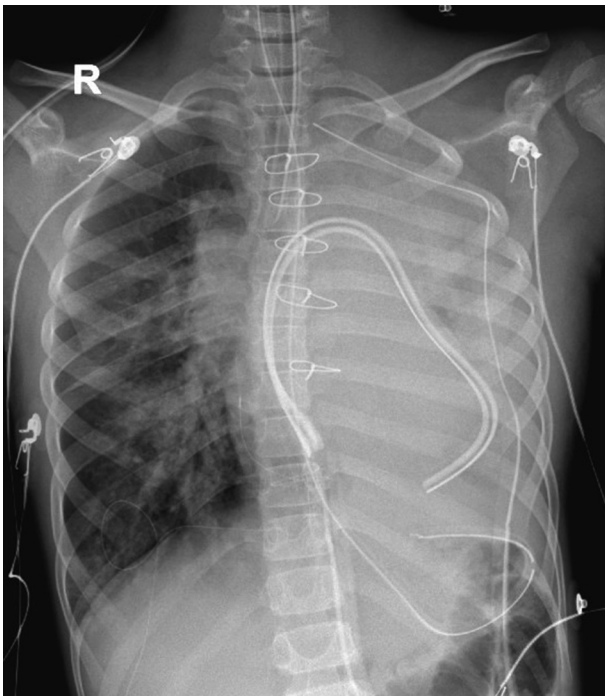


FIGURE E1. Radiograph of the chest demonstrating complete opacification of left lung, observed after unsuccessful attempts at extubation in the first few postoperative days.