Cardiology in the Young

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Fenestrated closure of an atrial septal defect for left ventricular diastolic dysfunction in an early infant with hypertrophic cardiomyopathy

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Brief Report

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Abstract

Left ventricular diastolic dysfunction is associated with poor prognosis in patients with hypertrophic cardiomyopathy and CHD. We report the case of an infant concomitant with hypertrophic cardiomyopathy, an atrial septal defect, and left ventricular diastolic dysfunction, who was successfully managed with fenestrated closure of the atrial septal defect.

Introduction

CHD occasionally accompanies hypertrophic cardiomyopathy. In such cases, complete closure of defects may reveal latent ventricular diastolic dysfunction. We report the case of an infant concomitant with hypertrophic cardiomyopathy, an atrial septal defect, and left ventricular diastolic dysfunction successfully managed with fenestrated atrial septal defect closure.

Case presentation

A 5-month-old girl was referred to our institution due to respiratory distress and convulsion followed by a respiratory tract infection. She has been diagnosed with hypertrophic cardiomyopathy by an echocardiogram and myocardial biopsy revealing hypertrophy of cardiomyocytes, interstitial fibrosis, and myocyte disarray (Supplementary Figure S1), and an atrial septal defect associated with cardiofaciocutaneous syndrome identified BRAF gene mutation. Chest radiography showed a cardiothoracic ratio of 0.68 with pulmonary congestion. Echocardiography revealed an interventricular septum thickness in diastole of 7.2 mm (+7.3 standard deviation), a left ventricular posterior wall thickness in diastole of 4.9 mm (+3.0 standard deviation), an interventricular septum thickness in diastole/left ventricular posterior wall thickness in diastole ratio>1.3 indicating asymmetric septal hypertrophy (Supplementary Figure S2), left ventricular ejection fraction of 68%, an atrial septal defect with 15 mm diameter, tricuspid pressure gradient increase up to 66 mmHg with central venous pressure of 15 mmHg measured directly using a central venous catheter, and no right ventricular outflow obstruction. Tissue Doppler echocardiography demonstrated an increase in the early diastolic transmitral velocity to early diastolic tissue velocity ratio. Subsequently, the patient was intubated, supported with mechanical ventilation, and treated with diuretics and antibiotics. However, echocardiography showed increased right ventricular and decreased left ventricular diameters. Inhaled nitric oxide caused a decrease in right ventricular systolic pressure and end-diastolic diameter (Figure 1). Cardiac catheterisation performed on the 18th day of admission showed a mean pulmonary arterial pressure of 18 mmHg and pulmonary to systemic blood flow ratio of 2.6 (Table 1). The patient underwent fenestrated atrial septal defect closure with a 4-mm fenestrated patch, with the suspicion that complete atrial septal defect closure might cause lung congestion due to left ventricular diastolic dysfunction. Postoperative cardiac catheterisation performed 7 months later showed the pulmonary to systemic flow ratio of 1.3; left ventricular diastolic dysfunction was considered latent because the balloon occlusion test of the atrial septal defect showed left ventricular diastolic pressure increase.

Discussion

This case highlights that an atrial septal defect with left ventricular diastolic dysfunction might require early intervention and that fenestrated atrial septal defect closure is a safe and effective treatment for such a condition.

Left ventricular diastolic dysfunction may necessitate early surgery for an atrial septal defect. The patient was diagnosed with hypertrophic cardiomyopathy, and left ventricular diastolic dysfunction was evidenced by an echocardiogram and confirmed through a closing test of the atrial septal defect in postoperative cardiac catheterisation. Right ventricular enlargement

Table 1. Preoperative and postoperative cardiac catheterisation data

	Preoperative data	Postoperative data	ASD closure test
RAP (mmHg)	4	4	
RVEDP (mmHg)	7	7	
mean PAP (mmHg)	18	21	
PCWP (mmHg)	5	6	
LAP (mmHg)	4	5	8
LVEDP (mmHg)	7	10	15
CI (L/min/m^2)	2.4	3.7	
Qp/Qs	2.6	1.3	
PVR (Wood unit · m^2)	2.1	3.3	
LVEDV (% of normal)	73.8	93.8	
LVEDVI (mL/m^2)	30	40.8	
RVEDV (% of normal)	172	140	

ASD = atrial septal defect; CI = cardiac index; LAP = left atrial pressure; LVEDP = left ventricular end-diastolic pressure; LVEDV = left ventricular end-diastolic volume; LVEDVI = indexed left ventricular end-diastolic volume; PAP = pulmonary artery pressure; PCWP = pulmonary arterial wedge pressure; PVR = pulmonary vascular resistance; RAP = right atrial pressure; RVEDP = right ventricular end-diastolic pressure; RVEDV = right ventricular end-diastolic volume.

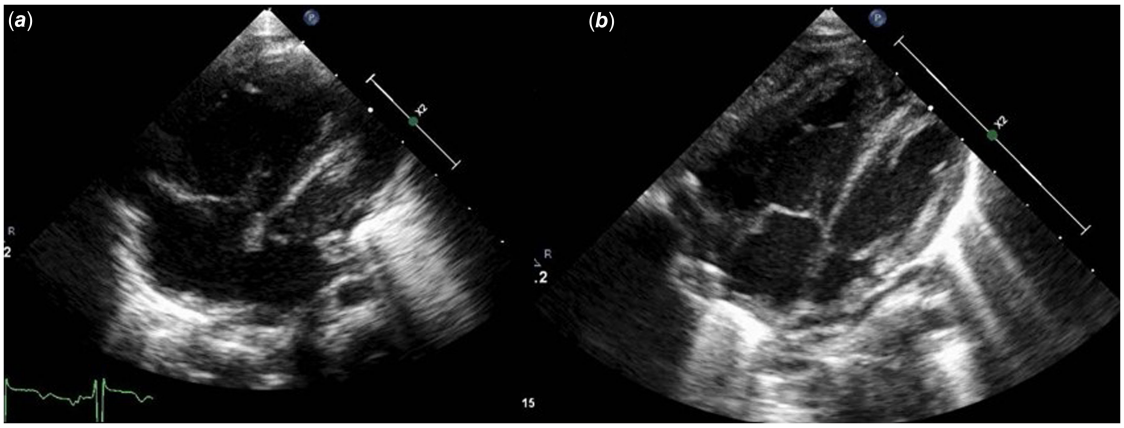

Figure 1. An echocardiogram showing (***a***) an enlarged right ventricle and (***b***) an enlargement of the left ventricle space after using inhaled nitric oxide.

related to a large atrial septal defect worsened due to pulmonary hypertension from lung congestion accompanied by convulsions and a respiratory tract infection. Inhaled nitric oxide treatment caused right ventricular shrinkage, left ventricular enlargement, and a dramatic increase in urine output. We hypothesised that the enlarged right ventricle pressed the left ventricle and worsened left ventricular diastolic dysfunction. The central venous pressure was 15 mmHg; however, a bidirectional shunt predominantly favoured left-to-right shunting across the atrial septum. Therefore, the inflow to the left ventricle decreased, and low cardiac output symptoms emerged. Neurogenic pulmonary oedema accompanied by convulsions is a cause of lung congestion. Although we used anti-epileptic drugs, the patient experienced recurrent seizures. Cardiofaciocutaneous syndrome is a RASopathy similar to Noonan syndrome but more frequently complicated with infantile spasms and neurodevelopmental delay.[1]

Giardini et al. showed that by closing an atrial septal defect, the right ventricle's volume overload decreased while the left ventricular diastolic dysfunction improved.[2] This suggests that ventricular interdependence is involved in ventricle diastolic dysfunction. In our case, inhaled nitric oxide improved pulmonary hypertension. The shrunken right ventricle released the left ventricle from suppression, increasing urine output and the left ventricular outlet tract's velocity time integral.

Complete closure of an atrial septal defect with left ventricular diastolic dysfunction may increase left atrial pressure, followed by lung congestion. To ensure that the atrial septal defect could be closed without complications, we needed to conduct a closing test. However, the patient's body was relatively small to insert an occlusion device for a large atrial septal defect preoperatively. In an adult case of atrial septal defect with hypertrophic cardiomyopathy, a closing test using a percutaneous closure device was conducted. If pulmonary artery pressure and pulmonary artery wedge pressure increased post-closure, a percutaneous fenestrated atrial septal defect occlusion device was selected, or diuretics, nitroglycerin, and vasoactive drugs were used.[3] Adaptation criteria for using a fenestrated occlusion device have not been definitively decided; however, if the left atrial pressure increased over 4 mmHg

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during a closing test[4] or exceeded 16 mmHg after a closing test,[5] a fenestrated device is recommended. We performed partial atrial septal defect closure using a fenestrated patch. The fenestration size was based on a report of left ventricular rehabilitation to a border zone left ventricular hypoplasia conducting atrial septal defect closure with a 4 mm fenestration.[6] Balloon dilation of the fenestration stenosis using an expanded polytetrafluoroethylene graft is reportedly effective in a post-fenestrated Fontan operation[7]; we performed partial atrial septal defect closure using an expanded polytetrafluoroethylene patch anticipating that balloon dilation might be necessary.

In our case, postoperative cardiac catheterisation revealed reduced pulmonary flow and low left atrial pressure. The reported risk factors for lung congestion post-atrial septal defect closure include a left atrial pressure of>10 mmHg pre-closure[8] or left ventricular end-diastolic pressure of>10 mmHg post-closure.[9] In our case, the left atrial and left ventricular end-diastolic pressures were elevated during the closing test, making complete atrial septal defect closure risky. Paediatric cardiac catheterisation is usually performed under sedation, and evaluation using atrial septal defect occlusion alone is inaccurate. Volume load and abdominal compression are also recommended.[10] If pulmonary hypertension worsens and is unrelated to high pulmonary flow, balloon dilation to the fenestration of an atrial septal defect would be needed, considering the left ventricular diastolic dysfunction involvement.

Conclusion

Atrial septal defects with left ventricular diastolic dysfunction may require early treatment, and fenestrated atrial septal defect closure is safe and effective in these cases.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951124036102.

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Competing interests. The authors declare none.

Ethical standards. This case report does not involve human and/or animal experimentation. The study protocol was approved by the Ethics Committee of Kobe Children's Hospital.

References

1. Pierpont MEM, Magoulas PL, Adi S, et al. Cardio-facio-cutaneous syndrome: clinical features, diagnosis, and management guidelines. Pediatrics 2014; 134 : 1149–1162.
2. Giardini A, Moore P, Brook M, Stratton V, Tacy T. Effect of transcatheter atrial septal defect closure in children on left ventricular diastolic function. Am J Cardiol 2005; 95 : 1255–1257.
3. Lim DS, Bergin JD, Ragosta M. Hypertrophic cardiomyopathy complicated by atrial septal defect and pulmonary hypertension. Catheter Cardiovasc Interv 2008; 71: 659–664.
4. Holzer R, Cao QL, Hijazi ZM. Closure of a moderately large atrial septal defect with a self-fabricated fenestrated Amplatzer septal occlude in an 85-year-old patient with reduced diastolic elasticity of the left ventricle. Catheter Cardiovasc Interv 2005; 64 : 513–518.
5. Abdelkarim A, Levi DS, Tran B, Ghobrial J, Aboulhosn J. Fenestrated transcatheter ASD closure in adults with diastolic dysfunction and/or pulmonary hypertension: Case series and review of the literature. Congenit Heart Dis 2016; 11: 663–671.
6. Emani SM, McElhinney DB, Tworetzky W, et al. Staged left ventricular recruitment after single-ventricle palliation in patients with borderline left heart hypoplasia. J Am Coll Cardiol 2012; 60: 1966–1974.
7. Kreutzer J, Lock JE, Jonas RA, Keane JF. Transcatheter fenestration dilation and/or creation in postoperative fontan patients. Am J Cardiol 1997; 79: 228–232.
8. Ewert P, Berger F, Nagdyman N, et al. Masked left bentricular restriction in elderly patients with atrial septal defects: a contraindication for closure? Catheter Cardiovasc Interv 2001; 52: 177–189.
9. Schubert S, Peters B, Abdul-Khaliq H, Nagdyman N, Lange PE, Ewert P. Left ventricular conditioning in the elderly patient to prevent congestive heart failure after transcatheter closure of atrial septal defect. Catheter Cardiovasc Interv 2005; 64: 333–337.
10. Satoshi M, Hideaki S. Diastolic dysfunction in congenital heart disease: clinical impact and basic evaluation. Pediatric Cardiology and Cardiac Surgery 2016; 32: 277–290.