Left ventricular restrictive physiology in kids with atrial septal defects: Something unexpected!

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ABSTRACT

Atrial septal defect (ASD) associated with left ventricular (LV) diastolic dysfunction (DD) is rare in children. DD is common in elderly patients undergoing ASD closure. Restrictive physiology among children undergoing percutaneous ASD closure is something unexpected that has not been described before in the literature. Hence, we report a child referred to our center for ASD closure in whom an LV DD has been unmasked with a balloon occlusion test and has been managed with pharmacological preconditioning and subsequent ASD closure with good outcome. Albeit rare, LV DD can occur in children undergoing ASD closure.

Keywords: Angioplasty, fenestration, interventional cardiology, percutaneous atrial septal defect closure

INTRODUCTION

Atrial septal defect (ASD) associated with left ventricular (LV) diastolic dysfunction (DD) is quite common in elderly patients undergoing ASD closure.[1] Facing children undergoing percutaneous ASD closure with a restrictive physiology is something unexpected and an exceedingly rare condition. Here, we report a case of a child referred to our center for ASD closure where an LV restrictive physiology has been unmasked with balloon occlusion test and where pharmacological preconditioning was successfully undertaken before transcatheter ASD closure.

CASE REPORT

A 3-year-old boy, weighing 16.2 kg, was regularly followed in outpatient clinic for secundum ASD diagnosed on echocardiogram done at birth for respiratory distress. The echocardiogram showed high pulmonary arterial pressure and signs of volume overload of right ventricle that increased over time, with a systolic pulmonary arterial pressure (sPAP) that rose from 35 to 45 mmHg. Therefore, he was referred to our center for a right and left heart catheterization. The electrocardiogram showed sinus rhythm with incomplete right bundle branch block. The echocardiogram was remarkable for a large secundum ASD (15 mm × 18 mm), with left-to-right shunt, right ventricular overload, mild mitral regurgitation and mild tricuspid regurgitation with a sPAP of 50 mmHg, normal biventricular function, no evidence of LV DD with a normal mitral inflow pulse wave Doppler, and a nondilated left atrium. Moreover, a normal coronary origin was identified in parasternal views.

The right heart catheterization showed a large hemodynamically significant secundum ASD with basal Qp/Qs of 1.8/1, mean PAP (mPAP) of 30 mmHg, and pulmonary vascular resistance of 2.8 WU.m^2. After oxygen

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delivery, a decrease in mPAP (22 mmHg) and an increase in Qp/Qs (5.5/1) were recorded. Unexpectedly, the left heart catheterization exhibited augmented LV end-diastolic pressure (LVEDP, 20 mmHg) and pulmonary capillary wedge pressure (18 mmHg). Normal systemic blood pressure was documented. We hence decided to hold the procedure and discharge the child home on diuretics and ACE inhibitors. Three months later, he was admitted again to perform a cardiac magnetic resonance and possible ASD closure. Magnetic resonance did not reveal any pathologic features of the LV: It had normal dimensions and mass, with no late gadolinium enhancement. Therefore, a new cardiac catheterization was undertaken. It confirmed the hemodynamic impact of the ASD (Qp/Qs of 2.5/1) with an mPAP of 14 mmHg. The LVEDP was slightly decreased from the first catheterization but remained high (14 mmHg), increasing up to 21 mmHg during complete inflation of an AGA sizing balloon 18 mm (Abbott Park Road Abbott Park, Illinois, USA) across the ASD [Figure 1a and b]. The stop flow was reached at 16 mm of balloon diameter. Considering both the hemodynamic significance of the ASD and the increased LVEDP, we decided to close the ASD creating two fenestrations in a 16-mm Amplatzer Septal Occluder (St. Jude Medical, St. Paul, MN, USA) inflating an 8-mm AndraBalloon (Andramed GmbH, Reutlingen, Germany) in two holes created using a 12-French Cordis catheter dilator (Cordis, Hialeah, Florida, USA). After release, the device was distorting the aortic root. Therefore, we decided to remove it because of risk of erosion. We retrieved it by using a 25-mm Amplatz Goose Neck Snare (Medtronic, Dublin, Ireland). Then, we implanted a 30-mm GORE CARDIOFORM septal occluder (GCSO; Goremedical, W. L. Gore and Associates, Inc., Newark, DE, USA) creating two fenestrations [Figure 2a and b]. The device did not seem to deform or compress the aortic root, so we released it. At the end of the procedure, the transesophageal echocardiogram showed a well-positioned device, with a mild left-to-right shunt across the fenestrations [Figure 2c]. An LVEDP of 20 mmHg and an mPAP of 21 mmHg were sampled at the cardiac catheterization [Figure 1c]. During the 4 days of postprocedural monitoring, the patient remained stable and asymptomatic. He was discharged on ACE inhibitors and acetylsalicylic acid. At 2-month follow-up, the result was stable and the patient was in NYHA Class I. At the transthoracic echocardiogram, the fenestration was not clearly visible, considering the diagnostic limits.

DISCUSSION

In patients with an overt DD, ASD can be seen as a pop-off valve that prevents substantial rise in pulmonary circulation pressure consequent to an increase in left atrial pressure (LAP). Therefore, before ASD closure, a complete left and right heart catheterization is strongly recommended in such patients. If a baseline elevation in LAP (>15 mmHg) or in LVEDP (>20 mmHg) is found, closure should not be performed without further assessment or intervention, because of the higher risk of pulmonary edema.[2] If basal LAP and LVEDP are normal, a balloon occlusion test with hemodynamic monitoring is suggested, due to the risk of a masked LV DD. In case of positive balloon occlusion test (increase >10 mmHg of LA or LVEDP and fall of systemic pressure),[1] physicians

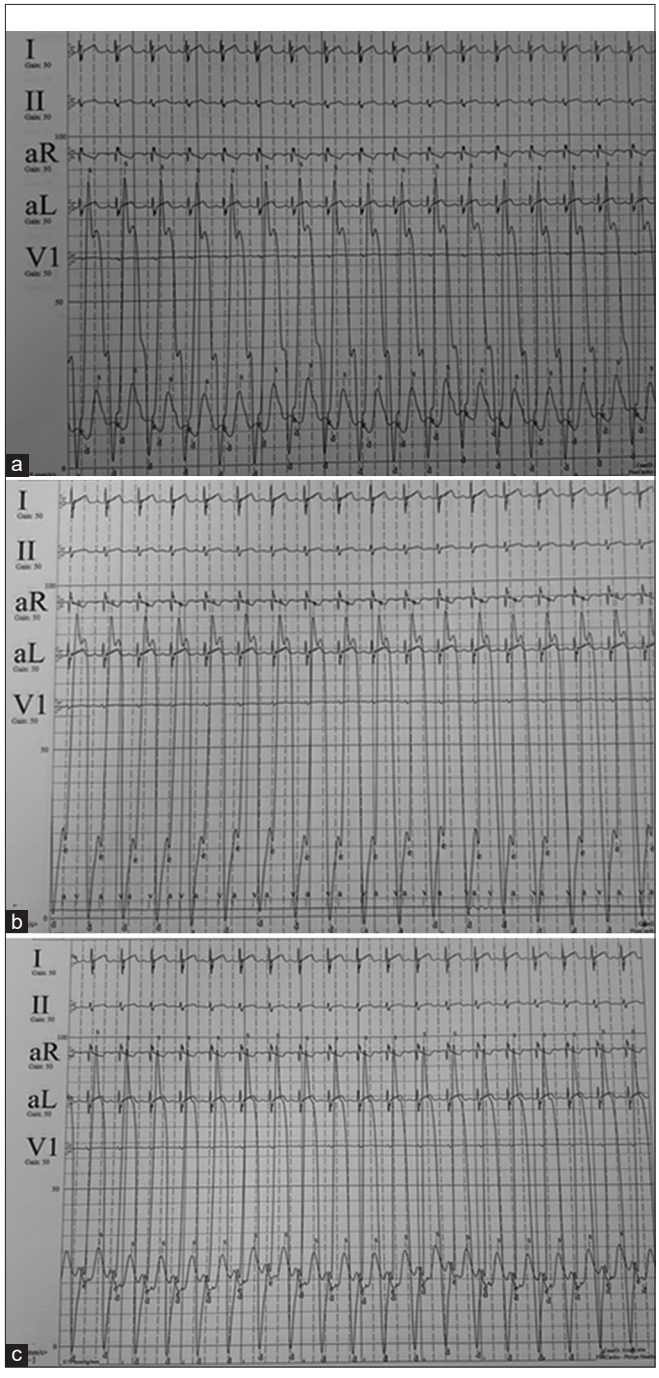

Figure 1: (a) Increased basal left ventricular end-diastolic pressure: 14 mmHg. (b) Further increase in left ventricular end-diastolic pressure at the balloon occlusion test: 21 mmHg. (c) Left ventricular end-diastolic pressure after fenestrated Gore Septal Occluder (GSO) implantation: 20 mmHg

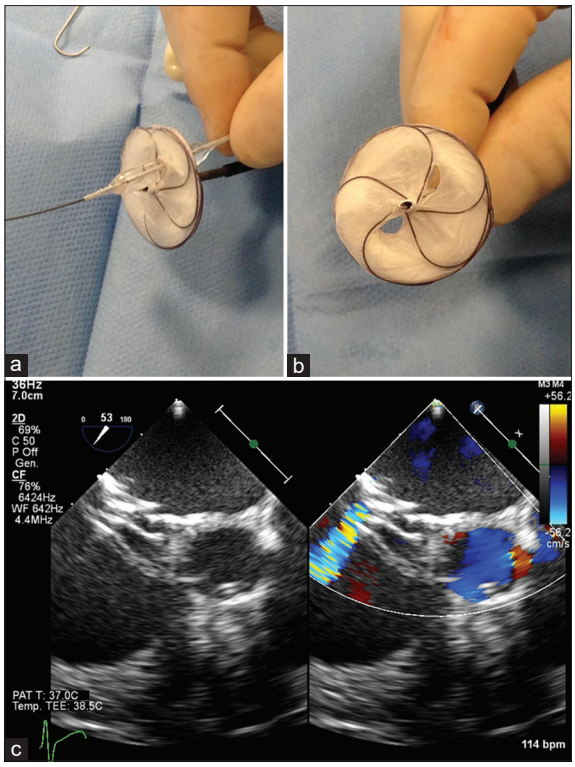

Figure 2: (a) 8-mm AndraBalloon inflated across GSO device to create a fenestration. (b) Two fenestrations created in a GSO device. (c) Left-to-right shunt across the GSO after its implantation

should first look for reversible causes of DD, such as myocardial ischemia or uncontrolled hypertension, which must be treated first. If a reversible cause is not found, such patients should be prepared with intravenous diuretics, vasodilators, and inotropes to face the abrupt increase of LVEDP after ASD closure.[1] Moreover, Gruner *et al.* showed that pharmacological preconditioning with ACE inhibitors or angiotensin receptor blockers plus oral diuretics before ASD closure may prevent periprocedural pulmonary edema.[3] Finally, in this subset of patients, the procedure can also be performed safely using a fenestrated closure device: It can be self-fabricated by creating a 4–6 mm fenestration to permit a residual shunting following defect closure.[4,5]

To the best of our knowledge, the association of ASD and LV DD has been very rarely described before in children. The evaluation and grading of DD in children are challenging and pediatric cardiologists have to rely on guidelines derived from studies on adult patients, which usually combines different echocardiographic parameters, such as mitral E/A ratio, mitral E/e' ratio, changes in mitral inflow with Valsalva maneuver, LA maximum volume index, pulmonary vein S/D velocity ratio, Ar-A duration, and TV regurgitation jet velocity. Moreover, left atrial and LV longitudinal strain has been identified as possible indicators of DD.[6] Hence, the prevalence of this condition may be underestimated in the pediatric population.[7] Furthermore, the presence of a pop-off mechanism, as an ASD, may mask the presence of a restrictive physiology as it happens also in the adult setting.[1,2,5]

CONCLUSION

ASD associated with LV DD is very rare in children. When encountered, it can be dealt similarly to adults with diuretics and ACE inhibitors for 6 months before ASD closure. A fenestrated device is also a valuable option.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. 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;64:333-7.
2. Tadros VX, Asgar AW. Atrial septal defect closure with left ventricular dysfunction. EuroIntervention 2016;12 Suppl X: X13-X7.
3. Gruner C, Akkaya E, Kretschmar O, Roffi M, Corti R, Jenni R, *et al.* Pharmacologic preconditioning therapy prior to atrial septal defect closure in patients at high risk for acute pulmonary edema. J Interv Cardiol 2012;25:505-12.
4. Kretschmar O, Sglimbea A, Corti R, Knirsch W. Shunt reduction with a fenestrated Amplatzer device. Catheter Cardiovasc Interv 2010;76:564-71.
5. Santoro G, Giugno L, Capogrosso C, Gaio G, Russo MG. ASD closure in special situations: Elderly, PA-IVS. In: Butera G, Chessa M, Eicken A, Thomson J, editors. Cardiac Catheterization for Congenital Heart Disease: From Fetal Life to Adulthood. Springer/Verlag Italy; 2015. p. 561-70.
6. Nagueh SF. Non-invasive assessment of left ventricular filling pressure. Eur J Heart Fail 2018;20:38-48.
7. Mawad W, Friedberg MK. The continuing challenge of evaluating diastolic function by echocardiography in children: Developing concepts and newer modalities. Curr Opin Cardiol 2017;32:93-100.