Balloon Valvuloplasty to Treat Adult Symptomatic Pulmonary Valve Stenosis with Sequential Follow-Up Using Cardiac Magnetic Resonance Imaging in Combination with Echocardiography

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Summary

Pulmonary valve stenosis (PVS) accounts for approximately 10% of all congenital heart defects. Echocardiography and right heart catheterization are the gold standards for diagnosis of PVS and for assessing disease severity and responsiveness to treatment.

Recently, cardiac magnetic resonance imaging (cMRI) has been established as an important tool to comprehensively evaluate cardiac structure and function; however, research into the usefulness of cMRI for PVS management is limited. Here, we describe a case of a 59-year-old female with isolated, severe PVS who was successfully treated with balloon pulmonary valvuloplasty (BPV) followed by sequential cMRI at 1 and 12 months. Exertional dyspnea and elevated plasma BNP concentration were observed 1 month after BPV; however, echocardiographic findings did not indicate recurrent stenosis or increased pulmonary valve regurgitation but an increase in mitral E/e'. cMRI demonstrated improved systolic forward flow and RV function with enlargement of LV volume, and the rapid increase in LV preload might be associated with the transient deterioration in symptoms and BNP level, which both gradually improved within 3 months after BPV. CMRI further depicted that a reduced RV mass index and increased RV cardiac output were achieved gradually during the follow-up period.

In conclusion, cMRI in combination with echocardiography was sufficiently informative to follow-up this PVS patient both before and after BPV. cMRI is easily reproducible in adult patients; therefore, cMRI should be recommended for long-term follow-up in adult PVS patients.

Key words: Congenital heart defect, Right ventricular function

Pulmonary valve stenosis (PVS) usually originates during fetal development as a result of improper formation of the pulmonary valve. It is associated with many congenital heart defects (CHDs). Moderate and severe cases of PVS may worsen and require surgery. Balloon pulmonary valvuloplasty (BPV) is the treatment choice for patients with symptomatic PVS, and echocardiography and right heart catheterization (RHC) are examination standards for the diagnosis, assessment of disease severity, and responsiveness to treatment in patients with PVS.

Recently, cardiac magnetic resonance imaging (cMRI) has been deemed an important tool for the comprehensive evaluation of various aspects of cardiac structure and function in patients with valvular or CHDs; however, research into the usefulness of cMRI for the management of isolated PVS is limited, even though PVS accounts for approximately 10% of all CHDs. Here, we describe a case of a 59-year-old female with isolated symptomatic PVS who was successfully treated with BPV and whose course of improvement was successively monitored using cMRI in combination with echocardiography.

Case Report

The patient was a 59-year-old Japanese female who had lived with a heart murmur since childhood. She was diagnosed with PVS at the age of 14 years, but was asymptomatic until her mid-50s. The patient then developed progressive dyspnea (New York Heart Association [NYHA] functional capacity class III), and was referred to our university hospital. She did not have peripheral edema, ascites or liver damage, however, she presented with a grade 3/6 systolic murmur at the third left sternal border, and enlarged left pulmonary arteries (PA) were identified on a chest roentgenogram. Electrocardiography
whereas RV cardiac output decreased to 1.71 L/minute/m². Ejection fraction (RVEF) was preserved at a value of 67%, detected. The RV chamber was not dilated, and the RV gadolinium enhancement; however, other CHDs were not 31.5 g/m² and muscular obstruction of the RVOT with late.

The patient was diagnosed with isolated, severe PVS with mild RVOT obstruction, and our heart team highly recommended treatment with BPV.

On cardiac catheterization, the RV systolic pressure (RVSP) was 104 mmHg, and the peak systolic pressure gradient between the PA and the RV was 84 mmHg. The RV end-diastolic pressure and mean right atrial pressure were within the normal ranges; 7 and 5 mmHg, respectively. Cardiac index and pulmonary capillary wedge pressure were 1.57 L/minute/m² (Fick) and 6 mmHg, respectively. Right ventriculogram revealed RVOT obstruction with a pulmonary valve annulus of 18 mm and post-stenotic dilation of the main PA (Figure A). We carried out transfemoral BPV to relieve the stenosis and improve cardiac performance. Two 12 × 25 mm Tyshak balloon catheters (B. Braun Interventional Systems Inc.) were positioned across the pulmonary valve and inflated simultaneously until the indentation of the balloon disappeared (Figures B, C). After successful dilatation, RVSP and the peak systolic pressure gradient drastically decreased to 51 and 37 mmHg, respectively (Table I). Echocardiography also revealed a decrease in peak systolic velocity to 3.0 m/second (pressure gradient = 36 mmHg) without pulmonary valve regurgitation (PR); therefore, the RV-PA pressure gradient was mainly due to PVS, and the additional treatment for RVOT obstruction was not required. Cardiac index (1.45 L/minute/m²) did not change just after BPV.

One month after treatment, echocardiography showed no recurrence of PVS; however, the patient still presented with mild exertional dyspnea, an increase in mitral E/e’ (9.7) detected by echocardiography, and an elevated plasma BNP concentration (83 pg/mL). She underwent plain cMRI because of nausea after the exposure to gadolinium contrast media at the first cMRI, and the cMRI demonstrated enlargement of the left ventricle (LV) with an LV end-diastolic volume from 59 and 82 mL, reductions in RV end-systolic volume from 22 to 13 mL and RV mass index from 31.5 to 25.7 g/m², and an increase in RVEF from 67% to 81% and RV cardiac index from 1.71 to 2.28 L/minute/m² (Table II). These observations suggested that RV systolic hypertension remained decreased after BPV with an improvement in systolic forward flow. The patient’s symptoms disappeared 3 months later (NYHA functional capacity class I). Follow-up evaluations carried out 12 months after BPV revealed a significant reduction in plasma BNP concentration (23 pg/mL), a decrease in mitral E/e’ (9.1), and right atrial enlargement. On cMRI, the RV cardiac index and RV mass index further improved to 2.66 L/minute/m² and 24.4 g/m².

Table I. Changes in Hemodynamic Parameters of Right Heart Catheterization Before and Just After BPV

| Parameter                                 | Pre  | Post |
|-------------------------------------------|------|------|
| Systolic right ventricular pressure (mmHg) | 104  | 51   |
| Mean pulmonary arterial pressure (mmHg)   | 12   | 13   |
| Pulmonary valve pressure gradient (mmHg)  | 84   | 37   |
| Pulmonary capillary wedge pressure (mmHg) | 6    | 5    |
| Cardiac output (L/minute)                 | 2.38 | 2.20 |
| Cardiac index (L/minute/m²)               | 1.57 | 1.45 |
Table II. Clinical Parameters and Treatment Outcomes

|                       | Pre   | At 1 month | At 12 months |
|-----------------------|-------|------------|--------------|
| Brain natriuretic peptide (pg/mL) | 50.7  | 83         | 23           |
| Echocardiography       |       |            |              |
| Pulmonary valve peak velocity (m/second) | 4.4   | 2.56       | 2.66         |
| Pulmonary valve peak gradient (mmHg)   | 78    | 26         | 28           |
| Right ventricular hypertrophy    | +     | +          | +            |
| Right atrial dilatation      | +     | +          | −            |
| Tricuspid valve regurgitation | trivial | trivial    | trivial      |
| Pulmonary valve regurgitation | trivial | trivial    | trivial      |
| Mitral E/A                | 1     | 1.1        | 0.7          |
| Mitral E/e'               | 9.4   | 9.7        | 9.1          |
| Cardiac MRI               |       |            |              |
| RVEDV (mL)               | 66    | 70         | 69           |
| RVESV (mL)               | 22    | 13         | 13           |
| RV EF (%)                | 67    | 81         | 81           |
| RV mass index (g/m²)      | 31.5  | 25.7       | 24.4         |
| RVCO (L/minute)          | 2.53  | 3.37       | 3.93         |
| RVCI (L/minute/m²)       | 1.71  | 2.28       | 2.66         |
| Pulmonary regurgitant fraction (%) | 9.0 | 4.1       | 5.5          |
| LVEDV (mL)               | 59    | 82         | 77           |
| LVESV (mL)               | 21    | 31         | 27           |
| LVEF (%)                 | 65    | 62         | 65           |

RVEDV indicates right ventricular diastolic volume; RVESV, right ventricular systolic volume; RV EF, right ventricular ejection fraction; RVCO, right ventricular cardiac output; RVCI, right ventricular cardiac index; LVEDV, left ventricular diastolic volume; LVESV, left ventricular systolic volume; and LVEF, left ventricular ejection fraction.

Discussion

The present report describes a case of adult isolated, severe PVS that was successfully treated with BPV and followed up with sequential cMRI at 1 and 12 months. cMRI provides a comprehensive evaluation of cardiac anatomy, function, and myocardial tissue characterization, however, few reports have described sequential cMRI follow-up results in patients who have undergone BPV to treat isolated PVS, because most previously reported cases of PVS treated with BPV were focused on the treatment of children, who were followed up by full echocardiographic evaluations. In the present case, cMRI demonstrated dramatic changes in cardiac chamber size and function after BPV, which was consistent with prior results using echocardiography and RHC. These observations reveal that cMRI is an appropriate modality to achieve a non-invasive follow-up evaluation of PVS both before and after BPV.

PVS occurs in isolation in approximately 10% of CHDs but is often associated with other congenital heart defects. BPV provides both acute- and intermediate-term gradient relief in patients with isolated PVS, and BPV is recommended for treatment of symptomatic patients with moderate or severe PVS severe PVS (peak pressure gradient > 64 mmHg) if RV function is normal. A combined evaluation utilizing echocardiography and RHC is the most common assessment tool for PVS and has revealed that a considerable number of patients had suboptimal results that required reintervention and/or surgical repair (e.g., patients presented with at least moderate PR during median follow-up after BPV). Voet, et al reported that 9.4% of patients who underwent BPV required reintervention, mostly for restenosis, with a median follow-up of 6.0 years (range, 0-21 years). According to a report by Devanagondi, et al, up to 60% of patients had moderate PR after BPV with a median follow-up of 15.1 years (range, 10.1-26.3 years). Thus, it is necessary to conduct a safe and effective long-term follow-up study.

Although echocardiography is the first-line cardiovascular imaging modality for the assessment of patients with CHD, there are some limitations, such as operator dependence and the need for adequate acoustic windows. cMRI is preferred because of the unrestricted access to cardiovascular anatomy and function, good reproducibility and versatility. The safety and imaging quality of non-invasive cMRI in particular have been recently confirmed in the evaluation of RV function in valvular disease or CHDs. Although some investigators have reported the long-term outcomes of PR severity and RV dilation and dysfunction after BPV (as calculated by cMRI), to the best of our knowledge, no reports have used sequential cMRI to monitor hemodynamics or RV function in patients with isolated PVS who underwent BPV. In the present case, cMRI was performed before BPV, and 1 and 12 months after BPV, which was successfully carried out and revealed gradual improvement of RV function. The patient’s RV cardiac index remained unchanged just after BPV, which is consistent with previous reports. Exertional dyspnea and elevated plasma BNP concentration were observed 1 month after BPV; however, there was no indication of recurrent stenosis or increased PR flow when examined using echocardiography. Importantly, echocardiography showed an increase in E/e’ and cMRI demonstrated improved systolic forward flow and RV function with enlargement of LV volume (Table II), which indi-
cated the rapid increase in LV preload might be associated with the transient deterioration in symptoms and plasma BNP concentration, and all of them gradually improved within 3 months after BPV. In the present case, cMRI evaluation 1 month after BPV was helpful to assess the patient’s cardiac performance on an outpatient basis.

Conclusions

We assessed an adult patient with isolated symptomatic PVS that was successfully treated with BPV and followed up with sequential cMRI for 12 months. Non-invasive cMRI in combination with echocardiography was sufficiently informative to follow up the patient both before and after BPV. cMRI is easily reproducible; therefore, the evaluation with combination of echocardiography and cMRI should be commended for long-term follow-up of PVS after BPV especially in adult patients.

Disclosure

Conflicts of interest: None.

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