

Pulmonary Valve Stenosis Combined with Patent Foramen Ovale in Adult Patients: To Close or Not?

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Abstract

Pulmonary valve stenosis is one of the most common congenital heart diseases in adults as well as patent foramen ovale. Pulmonary valve stenosis combined with patent foramen ovale in adults is considered not rare in heart diseases in adults, although there is a lack of published evidence so far. This report outlines the management of pulmonary valve stenosis combined with patent foramen ovale in adults.

Keywords: Pulmonary valve stenosis; Patent foramen ovale; Cardiopulmonary bypass

Introduction

Pulmonary valve stenosis (PVS) is a heart valve disorder in which outflow of blood from the right ventricle of the heart is obstructed at the level of the pulmonic valve. This type of pulmonic stenosis results in the reduction of flow of blood to the lungs. These fade very rapidly in most children but they will not go away altogether. Smaller scars on the hands and neck usually fade away to nothing. One problem that can happen after a procedure or later on is that the pulmonary valve begins to leak. If this happens, it may need further repair or replacement.

Case Report

A 60-year-old woman (43.5 kg, 157 cm, BMI 17.6) was admitted to hospital. She presented with increasing dyspnea on exertion and cyanosis after intense activity from birth. These symptoms worsened for recent 4 months with edema of lower extremity. On physical examination, she was found a heart rate of 74 beats/min, respiratory rate of 20 breaths/min, blood pressure of 100/61 mmHg, temperature of 36.8°C.

Cardiovascular exam revealed a normal rate and rhythm, but a grade 2/6 holosystolic murmur was heard at the left sternal edge in the second intercostal space consistent with pulmonary stenosis. In non-oxygen state, percutaneous oxygen saturation was 82.7% and hemoglobin was 173 g/L. Transthoracic echocardiography revealed pulmonary stenosis (severe) and congenital patent foramen ovale with 114 mmHg maximum pressure across the pulmonary valve and bidirectional shunt through the gap between the foramen ovale. Clinical diagnosis was pulmonary stenosis (severe), congenital patent foramen ovale. Because of guide wire failed to reach the pulmonary artery via right ventricular outflow tract, interventional operation was abandoned and changed to cardiothoracic surgery.

The patient was taken to the operating room to have pulmonary valve replacement, right ventricular outflow tract reconstruction and closure of the patent foramen ovale under cardiopulmonary bypass. Once stop the extracorporeal circulation devices, right atrium was found swollen. Arterial blood gas was measured as pH 7.4, pCO₂ 40.7 mmHg, pO₂ 66.9 mmHg, oxygen saturation 90.6%. The disturbance of blood circulation had taken into account. After debating the pros and cons, the patient was heparin once again to reopening the foramen ovale.

After returning to cardiac surgery ICU, conventional electrocardiographic monitoring and mechanical ventilation were

used. Diuretics, vasoactive drugs and cardiotoxic drug were used to insure effective circulating blood volume. Sildenafil was administered 50 mg/d combined with Bosentan 125 mg /d to decrease pulmonary artery pressure. Arterial blood gas was measured as pH 7.45, pCO₂ 34.3 mmHg, pO₂ 63.7 mmHg, oxygen saturation 94.6% 3 days after operation. Transthoracic echocardiography revealed pulmonary transvalvular pressure was about 22 mmHg with a remarkable improvement. Bidirectional shunt through the PFO was seen, mainly left to right. After six-month follow-up, the patient recovered well. Oxygen saturation of the patients increased to 100%. No hypoxemia nor pulmonary hypertension were observed.

Discussion

The pathophysiology of this case is that due to longer duration of preoperative pulmonary valve stenosis, right ventricular ejection resistance increased and right atrial pressure elevated lead to increase in right ventricular load, causing right ventricular hypertrophy, decreased ventricular wall compliance and dysfunction of right ventricular. This mainly relies on right-to-left shunt through the foramen ovale to reduce the right heart load. On concern of the age, inadequate pulmonary vascular bed perfusion last from birth for 60 years thus has turned out to be dysplasia in pulmonary vascular bed. After debridement of right ventricular outflow tract, pulmonary valve stenosis was relieved. However, when large amounts of blood which discharged from relief of pulmonary valve stenosis enter dysplastic pulmonary vessels, it would lead to pulmonary hypertension. With intraoperative suture of foramen ovale, blood in right ventricular could not flow out and lead to a significant increase in right heart load eventually. Under such circumstances, foramen ovale was reopening to reduce the right heart load especially right atrial load. And by treating pulmonary hypertension postoperative, the patient recovered well.

So, the point is to identify patients who would benefit from closure of PFO in this situation. Right-to-left shunt caused by patent foramen ovale concurrent with pulmonic stenosis is reported in dog experiment,

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PFO with R-L shunt was more common with very severe Pulmonic Stenosis and absent with mild pulmonic Stenosis [1]. It has been reported that in children with pulmonary valve stenosis combined with PFO, for those with severe pulmonary valve stenosis and right ventricular failure, retaining the foramen ovale for about 4-9 mm in surgery can reduce right heart load, to facilitate right cardiac system development, otherwise easily lead to severe right heart failure [2]. And earlier repaired in life, the more improved prognosis. Postoperative treatment mainly centers on improve the function of right heart and lower pulmonary hypertension.

Conclusion

Treatment like inhaling NO, endothelin receptor antagonists like bosentan, phosphodiesterase type 5 inhibitor like sildenafil and prostanoids like epoprostenol are recommended to reduce pulmonary vascular resistance and improve the function of right heart [3]. Besides, phentolamine and captopril are reported to lead to vasodilation of the pulmonary and systemic vascularization, making right-to-left shunt across PFO decreased and saturation improved [4]. Moreover,

pulmonary arterial vasodilator drugs can be considered as treatments before surgery. Pulmonary blood flow indicators include McGoon ratio and pulmonary arterial index can provide information about pulmonary vessel development but lack large sample statistics so far. So still more data are needed to evaluate the benefits versus the risks of closing the PFO in patient concurrent with pulmonary valve stenosis.

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