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A Case of Severe Pulmonary Regurgitation Due to the Absence of Pulmonary Valve

Hyun-Ju Yoon, MD, Kye-Hun Kim, MD, Young-Keun Ahn, MD, Myung-Ho Jeong, MD, Jeong-Gwan Cho, MD, Jung-Chaee Kang, MD and Jong-Chun Park, MD

The Heart Center of Chonnam National University Hospital, University Research Institute of Medical Sciences, Gwangju, Korea

Tetralogy of Fallot with absent pulmonary valve is a very rare form of congenital heart disease with various clinical presentations. We experienced a 25-year-old female of severe pulmonary regurgitation due to absent pulmonary valve who had a history of open heart surgery for tetralogy of Fallot and review the literatures.

KEY WORDS : Pulmonary valve · Tetralogy of Fallot.

Introduction

Absent pulmonary valve is defined as total or subtotal absence of pulmonary valve leaflets. Stenosis of pulmonary artery orifice and aneurysmal dilatation of pulmonary arteries coexist in all cases. Absent pulmonary valve can be associated with simple and complex cardiovascular malformations.[1] Here, we report a rare case of severe pulmonary regurgitation (PR) caused by the absence of pulmonary valve in a 25-year-old female who had a history of tetralogy of Fallot (TOF).

Case

A 25-year-old female presented with dyspnea on exertion and leg edema which developed from 6 months ago. She had a history of corrective open heart surgery due to the TOF and hypoplastic left pulmonary artery (PA) 17 years ago in the U.S.A. Blood pressure was 90/60 mmHg, heart rate was 110 beats/min, and body temperature was 37°C. On physical examination, left parasternal heave and diastolic murmur along the mid-left sternal border were noted. Electrocardiography showed sinus rhythm and incomplete right bundle branch block. Chest X-ray showed cardiomegaly and prominence of main pulmonary artery (MPA). Laboratory findings were not significant. Trans-thoracic echocardiography (TTE) revealed enlarged right ventricle (RV), and severe pulmonary regurgitation (PR) due to the absence of pulmonary valve, but abnormal intracardiac shunt flow was not observed (Fig. 1). Evaluation of the heart and PA using multi-detector CT (MDCT) revealed marked enlargement of the RV and MPA. Three dimensional angioscopic reconstruction view of MDCT revealed dilated MPA, complete absence of pulmonary valve, and hypoplasia of the left PA (Fig. 2).

The patient was discharged with medication and waiting for the reconstruction surgery of pulmonary valve.

Discussion

Absent pulmonary valve syndrome is an uncommon form of congenital heart disease. It occurs in 2.4 to 6.3% of patients with TOF.[2] This syndrome was first reported in 1930.[3] TOF with absent pulmonary valve is a rare congenital anomaly characterized by features of TOF with either rudimentary ridges or the complete absence of pulmonary valve tissue. TOF consists of a malignant ventricular septal defect, infundibular pulmonary stenosis, overriding aorta, and right ventricular hypertrophy. The absence of mature pulmonary valve tissue leads to severe PR. This is often associated with massive dilatation of the pulmonary arteries, which is a characteristic of this syndrome.[4] The main symptoms are recurrent wheezes and dyspnea due to compression of the trachea and bronchi by the aneurysmal pulmonary arteries.

Isolated PR after TOF repair is not a common finding,

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• Address for Correspondence : Jong-Chun Park, The Heart Center of Chonnam National University Hospital, University Research Institute of Medical Sciences, 671 Jebongno, Dong-gu, Gwangju 501-757, Korea Tel : +82-62-220-6241, Fax : +82-62-225-6260, E-mail : jcpark54@hanmail.net

and the presence of lesions that further increase the RV afterload such as peripheral pulmonary artery stenosis could aggravate RV dilatation in addition to PR. In the additional presence of such lesions, PR is even less tolerated, and early pulmonary valve replacement is especially important to preserve RV function.[5]

Long term PR has been reported to cause RV dysfunction before overt clinical symptoms develop.[6] The abnormal characteristics of surgically repaired RV, such as conduction disturbances, resection of muscle, and hypertrophy, may add to the RV dysfunction. If not corrected, PR can lead to RV dilation, exercise intolerance, arrythmias and an increased incidence of sudden death.[7][8] So, surgical relief of the larger airway compression alone is not always effective in reversing the serve obstructive respiratory disease.

Optimal timing of pulmonary valve replacement is still a subject of debate. So far, clinical symptoms, exercise tolerance, ventricular dilatation, and onset of tricuspid regurgitation have been used as indicators for reintervention. In addition, in symptom free patients with right ventricular dilatation, pulmonary valve replacement has been considered for prevention and reduction of right ventricular dilatation. Additional arguments for this strategy are the predisposition of severe PR to ventricular dysrhythmias, and the beneficial effect of pulmonary valve replacement on electrical instability such as the incidence of tachyarrythmias.[9][10]

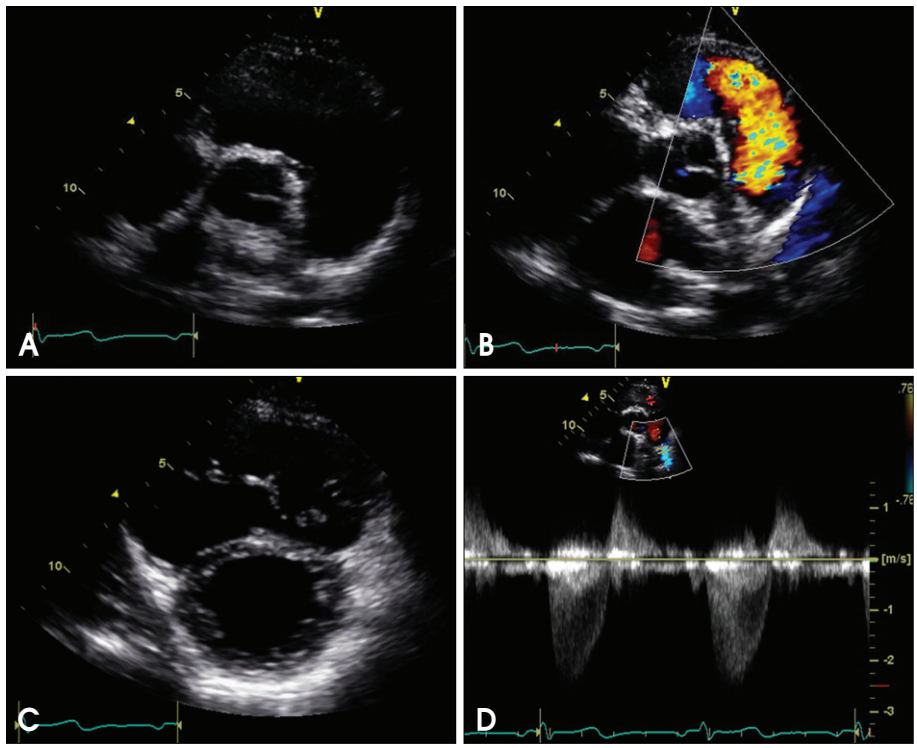

Fig. 1. Transthoracic echocardiogram revealed enlarged right ventricle, pulmonary artery, and severe pulmonary regurgitation associated with absence of pulmonary valve (A, B and C). Left pulmonary artery peak velocity was 2.5 m/s in parasternal short axis window (D).

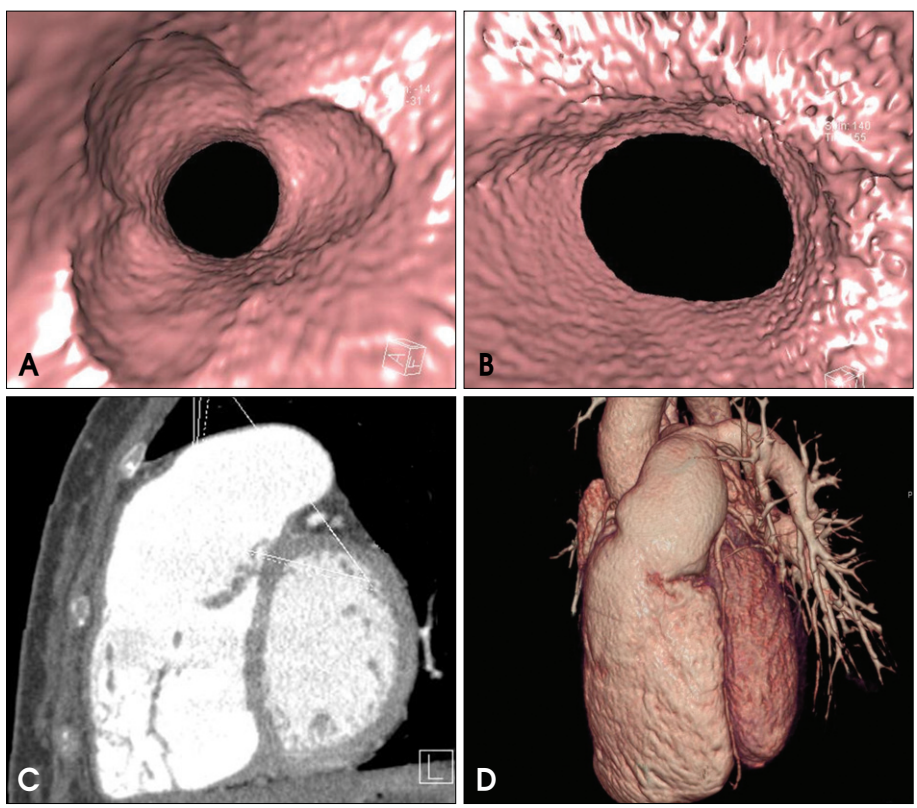

Fig. 2. MDCT with 3D-reconstruction showed three leaflets of pulmonary valve in normal person (A) and absence of pulmonary valve leaflets in this patient (B). CT revealed enlarged right ventricle and pulmonary artery (C and D).

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