Aneurysmally dilated RVOT in TOF with Absent Pulmonary Valve

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Title

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TOF and Absent Pulmonary Valve with Aneurysmal Right Ventricular Outflow Tract

In the present issue of the journal, Mishra and collogues have presented interesting images of an adult patient with Tetralogy of Fallot with absent pulmonary valve and aneurysmally dilated right ventricular outflow tract (1).

Survival of a patient diagnosed with Tetralogy of Fallot and absent pulmonary valve into adulthood, is a rarity. Patients diagnosed with this condition usually present in infancy with symptoms of respiratory distress and tachypnea, mainly because of the tracheobronchial compression resulting from massive dilatation of the main pulmonary artery and its first and second order branches. The consequential tracheomalacia and bronchomalacia determines the timing and severity of respiratory compromise.

This patient presented at the age of 37 years with symptoms of dyspnea without any history of cyanosis. Such a late presentation might have been due to the insignificant airway compression by the dilated main and branch pulmonary arteries. In such patients, the right ventricular infundibulum is aneurysmally dilated because of long standing pulmonary regurgitation and right ventricular outflow tract obstruction. Contrary to what its name suggests, there is presence of rudimentary and dysplastic pulmonary leaflets in this condition.

A one-stage repair is the preferred method of choice. Asymptomatic patients are usually operated on at 6-12 months of age and symptomatic patients are operated on a semi-urgent basis. Various surgical approaches have been described to correct this anomaly. The closure of ventricular septal defect, relief of right ventricular outflow tract obstruction, and achievement of a competent pulmonary valve are the primary goals of the surgery. These are followed by a specific aim to decompress the airways from dilated pulmonary arteries, if required. In adults, establishing competence of the pulmonary valve can be comparatively easy as compared to an infant. Reconstruction of the right ventricular outflow can be done using a bioprosthetic valve or hand sewn valve leaflets.

1. Mishra AK, Rana P, Raja J, Bansal V, Singhal M. Aneurysmal right ventricular outflow in an adult patient with unrepaired tetralogy of Fallot and an absent pulmonary valve. Journal of Cardiac Surgery. 2021. . . .