Corrected Tetralogy of Fallot in an Adult: A Case of Long-term Complications

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ABSTRACT

Introduction: Tetralogy of Fallot is a congenital heart defect consisting of a ventricular septal defect, overriding aorta, pulmonary artery stenosis and right ventricular hypertrophy. It is the most common cause of cyanotic congenital heart defect occurring in approximately 3.6 infants per 10,000 births and present in roughly 10% of congenital heart defect cases. Historically, more than 90-95% of patients with tetralogy of Fallot died before the age of 20 without surgery. However, with the advent of improved cardiovascular surgical techniques, more than 80% of these patients survive to adulthood, posing new challenges for the management of adult patients with tetralogy of Fallot.

Clinical Presentation: Patient AH was a 26-year-old male with a history of three corrective surgeries for tetralogy of Fallot who was seen due to complaint of random episodes of lightheadedness and syncope, accompanied by substernal chest pain. Imaging revealed a stenotic conduit and runs of non-sustained ventricular tachycardia.

Conclusion: The presentation and management of tetralogy of Fallot is historically discussed in children born with the congenital cardiac defect. However, as surgical interventions have become more successful in treating these cases, the focus needs to shift to include long-term management of the sequelae arising in adults following corrective surgery.

CASE PRESENTATION

• AH was a 26-year-old male with a history of corrected tetralogy of Fallot who was seen due to complaints of random episodes of lightheadedness and syncope, accompanied by substernal chest pain. He denied any known provocations. He had a history of three corrective surgeries: the first at 2 days old, then 2 months old and the last at 16 years old. Despite the corrective surgeries, he required supplemental oxygen over the last several weeks at variable times throughout the day and while sleeping at night due to occasional oxygen desaturation per his pulse oximeter and development of peripheral cyanosis.
• On physical exam, AH was a thin-appearing young man in no apparent distress. Cardiac exam revealed a holosystolic murmur located at the left sternal border. Lungs were clear to auscultation. There were no signs of peripheral or central cyanosis or clubbing of his digits.
• An EKG was performed in office to establish a baseline while a 30-day event monitor was ordered to be worn at home. He was also to follow up for a chest x-ray and a trans-thoracic echocardiogram.

DISCUSSION

Tetralogy of Fallot (TOF) is a congenital heart defect consisting of a ventricular septal defect (VSD), overriding aorta, pulmonary artery stenosis and right ventricular hypertrophy. It occurs when there is anterior and cephalad deviation of the infundibular septum resulting in a malaligned ventricular septal defect with an overriding aortic root that causes a right ventricular outflow tract (RVOT) obstruction. The degree of right ventricular outflow tract obstruction and thus adequacy of pulmonary flow is the most important determinant of prognosis. As obstruction increases, there is increased resistance of pulmonary blood flow, which causes a right-to-left shunt of blood flow, and an increase in the amount of deoxygenated blood bypassing pulmonary circulation to move directly into systemic circulation.

TOF surgery may include repair of the VSD and/or creating a conduit to increase the blood flowing into the pulmonary artery and thereby decreasing the rate of deoxygenated blood entering the aorta. The rate of TOF children surviving into adulthood has increased from less than 10% to close to 85% with the advent of the cardiothoracic repair surgery.

Postoperative TOF repair requires long-term follow up due to risk of developing chronic complications. In addition to creating a right bundle branch block, complications include pulmonary regurgitation, residual RVOT obstruction, right ventricular dysfunction, cardiac arrhythmias such as atrial valve insufficiency, and sudden cardiac death. Right ventricular dysfunction can lead to exercise intolerance and right heart failure, and other arrhythmias such as ventricular tachycardia, atrial flutter, and atrial fibrillation.

As this population of repaired TOF patients grow older, more sequelae will be discovered that needs to be monitored and managed.

IMAGING

30-Day event monitor: Runs of non-sustained ventricular tachycardia (VT)

Echocardiogram (not shown): TOF repair with stenotic right ventricle-pulmonary artery conduit with severe conduit regurgitation, right ventricular enlargement, and moderate tricuspid regurgitation. No residual ventricular defect or aortic regurgitation.

REFERENCES/ACKNOWLEDGEMENT

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