


Management of Unoperated Tetralogy of Fallot in a 59-Year-Old Patient

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Abstract

Tetralogy of Fallot is the most common cyanotic congenital heart defect consisting of an overriding aorta, right ventricular outflow obstruction, ventricular septal defect, and right ventricular hypertrophy. Without surgical management, approximately only 3% of patients survive past the age of 40 years. Cases of unoperated patients reaching adulthood have been reported; however, few studies describe treatment guidelines for surgical or therapeutic management. In this article, we report the case of a 59-year-old Hispanic male with unoperated tetralogy of Fallot presenting to our cardiology clinic for initial workup and management.

Keywords

tetralogy of Fallot, management, unoperated, unrepaired

Introduction

Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart disease affecting approximately 1660 infants in the United States each year.¹ First described by Etienne-Louis Fallot in 1888, ToF consists of right ventricular (RV) hypertrophy, a ventricular septal defect (VSD), RV outflow tract obstruction, and an overriding aorta.²

Surgical repair with the Blalock-Taussig procedure was initially described in 1945 with favorable long-term outcomes.^{3,4} Few patients with ToF survive into adulthood without operation; 10% surviving to their 30s and only 3% reaching their fourth decade.⁵ Controversy exists regarding the optimal age for repair. Typically, surgical correction of ToF is performed during infancy with the majority of centers aiming for complete correction by 9 months of age.⁶

Although infrequent, physicians may encounter patients who have never undergone attempted repair or palliative procedures. Without extensive documentation, it is difficult to determine the best course of treatment for such patients. We report the case of a 59-year-old Hispanic male with unrepaired ToF presenting to our cardiology clinic for management.

Case Report

A 59-year-old Hispanic male who recently emigrated from Mexico presented to our cardiology clinic with complex congenital heart disease consisting of unoperated ToF complicated by Eisenmenger syndrome and atrial fibrillation. The

patient was aware of his unrepaired congenital heart disease and followed by a cardiologist in Mexico. At the age of 19 years, he was advised to undergo surgical repair. However, he did not follow through due to fear of risks involved. He described experiencing lower extremity edema, dyspnea on exertion, and intermittent shortness of breath, often severe enough to limit him from speaking. He denies having any chest pain or orthopnea.

At his initial visit, the patient was normotensive (124/71 mm Hg), afebrile, with a normal respiratory rate, and 92% oxygen saturation on room air. His physical examination was noted to have an irregularly irregular heart rate and rhythm, a grade IV harsh crescendo systolic murmur throughout the precordium, grade I blowing diastolic murmur at the upper left sternal border with RV heave, and palpable thrill. Apical impulse was displaced inferolaterally. Chest radiography revealed cardiomegaly and prominence of the pulmonary vasculature (Figure 1).

Atrial fibrillation (82 beats per minute) was confirmed on electrocardiogram. Right and left heart catheterization concluded the patient had normal coronaries and severe pulmonary

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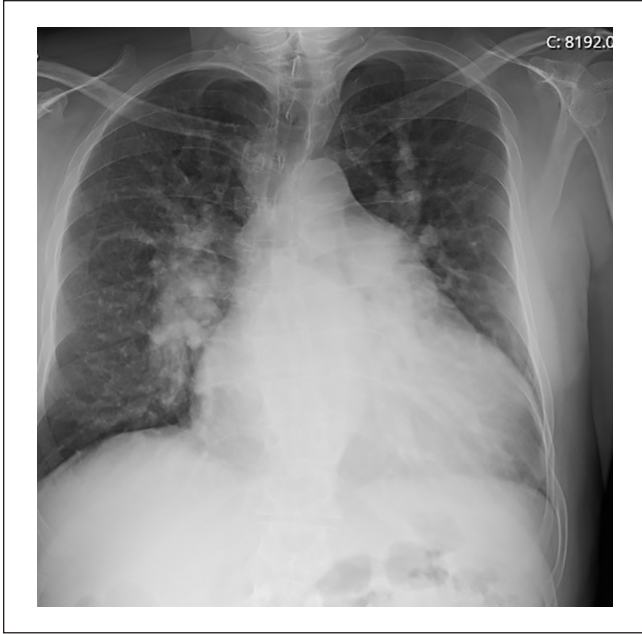


Figure 1. AP chest radiography of an unoperated tetralogy of Fallot adult patient revealing cardiomegaly and prominence of the pulmonary vasculature.

hypertension (HTN) evidenced by a mean pulmonary artery pressure of 53 mm Hg. No VSD flow was demonstrated through left ventricular (LV) angiogram; however, faint VSD flow was seen with RV angiogram consistent with Eisenmenger syndrome. RV systolic function was decreased and severely dilated. His LV ejection fraction was noted to be 45% to 50%.

Medical management was initiated with 4 mg warfarin twice a day (BID), sildenafil 20 mg (thrice a day) TID with a goal to increase to 40 mg TID, furosemide 40 mg BID, atorvastatin 20 mg daily, aspirin 81 mg daily, benazepril 10 mg daily, and metoprolol tartrate 25 mg BID.

Following treatment, the patient reported a decrease in lower extremity edema and shortness of breath. Repeat transthoracic echocardiography revealed dilated left and right ventricles with coexisting hypertrophy, moderate tricuspid and mitral regurgitation, severe pulmonary HTN, mild pulmonary stenosis with peak gradient of 36 mm Hg, low-velocity bidirectional shunting across the VSD, and a decreased ejection fraction 40% to 45%. He continues to be medically managed with adjustments to his regimen as needed. At his last visit, aspirin was discontinued and sildenafil had reached our targeted goal. Furosemide, metoprolol, and benazepril were all maintained at previous doses with warfarin monitored by our anticoagulation clinic. He is scheduled for repeat right heart catheterization in April 2020. A cardiothoracic surgery consultation is pending for further recommendation. He continues therapeutic management with the potential to bridge to surgical intervention.

Discussion

Our patient requires a multidisciplinary approach at a facility with adult congenital heart disease experts for late repair considerations of unoperated ToF. Albeit rare, cases of survival to middle age have been reported.

In a study of unoperated ToF patients at autopsy, survival without surgery was 66% at 1 year of age and 3% at 40 years.⁵ The first report of an uncorrected ToF patient surviving into late adulthood was made in 1929. Currently, an 87-year-old female is documented as the oldest known surviving patient.⁷

Turkish physicians describe a similar case of unoperated ToF in a 68-year-old woman with diabetes mellitus. This patient presented with similar symptoms of edema and dyspnea as well as a physical examination consistent with our patient. She declined surgical intervention.⁸ Unfortunately, her medical management and outcome was not further described. A 67-year-old man found to have unoperated ToF presented with cyanosis and clubbing. However, this patient had an uncommon coexisting manifestation of systemic HTN. Similarly, he declined surgical intervention. He was treated conservatively for congestive cardiac failure with furosemide, spironolactone, and ACE inhibitors for HTN. At his 3-month follow-up, he began carvedilol for heart failure, better hypertensive control, and decreased risk of sudden cardiac death with a stable course.⁹

Studies suggest that unoperated survivors have 3 common features: a hypoplastic pulmonary artery with moderately slow development of subpulmonary obstruction, LV hypertrophy (LVH), and systemic-pulmonary artery collaterals.¹⁰ Our patient had LVH consistent with other published cases. It is presumed that LVH may develop to delay right to left shunting and often occurs later in adulthood.¹⁰ No systemic-pulmonary artery collaterals were demonstrated in our patient. Anatomic detail, such as variant coronary anatomy, is of clinical significance when planning pre- and post-operatively.

Complete surgical repair has been performed since 1954 with early intervention recommended.³ Correction is now routinely performed with excellent results. Few studies describe the outcome or benefit of primary late surgical repair. Patients undergoing surgical repair at age 40 years or older were found to have improvement in functional class.² Reoperation was necessary in 14% of survivors with the most frequent cause being severe pulmonary regurgitation.² This emphasizes the importance of pulmonary valve replacement at initial operation. Mayo conducted a series of 30 patients undergoing total correction between the ages of 40 and 60 years. Their operative mortality was 3%. Postoperative long-term survival rate at 5 years and 10 years was 92% and 74%, respectively.¹¹ In relation to our patient, surgical repair should then be considered, as late operative intervention was associated with low mortality.

Although uncommon, physicians may encounter patients without having undergone any repair or palliation. Our patient highlights the importance of documenting medical and surgical management of the unoperated adult ToF patient to help establish therapeutic guidelines and recommendations.

Conclusion

Unrepaired ToF is of clinical significance when assessing late primary treatment options. Surgical repair should be considered, as late operative intervention is associated with low mortality. If procedural intervention is supported, it is important to note anatomic detail such as variant coronary anatomy for an optimal surgical outcome. Medical management is necessary for those who decline surgical intervention or are deemed poor candidates. Documenting additional cases of primary late repair of ToF is warranted to further guide treatment recommendations.

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Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Verbal informed consent was obtained from the patient for their anonymized information to be published in this article.

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