CLINICAL VIGNETTE

Pulmonary Regurgitation in Repaired Tetralogy of Fallot

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Introduction

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease, estimated at 1 in every 3600 live births. TOF is classically characterized by a tetrad of i) ventricular septal defect, ii) over-riding of the aorta, iii) right ventricular outflow obstruction, and iv) right ventricular hypertrophy. Primary corrective surgery is the treatment of choice and is generally performed in the first year of life. Goals of primary corrective surgery include closure of the ventricular septal defect and relief of right ventricular outflow obstruction, while preserving the right ventricular form and function and maintaining pulmonary valve competence.

With improving surgical techniques, the majority of patients with TOF are surviving into adulthood. However, late surgical complications, such as pulmonary valve incompetence, are increasingly recognized in these survivors. With the right ventricular outflow tract obstruction relieved during surgical repair for TOF, pulmonary regurgitation (PR) often develops, resulting in right ventricular dilatation and subsequent progressive exercise intolerance, right heart failure, arrhythmia, and sudden death. Diagnosis of this late surgical complication can be made by 2D and color Doppler echocardiogram and careful assessment of PR is important in determining the timing and approach to pulmonary valve replacement.

Case Summary

A 47-year-old male with a history of Tetralogy of Fallot status post-left-sided Blalock-Taussig shunt at age 7 months old and intra-cardiac surgical repair at age 7-years-old presented with two months of shortness of breath, frequent palpitations, intermittent chest pain, and one episode of syncope. Since his TOF repair during childhood, the patient maintained a normal lifestyle except for mild exercise intolerance. Vital signs were significant for a heart rate ranging from 100 to 120 bpm, blood pressure of 118/84, and oxygen saturation of 98% on room air. Physical exam was notable for a 3/6 holosystolic murmur and early 3/6 diastolic murmur loudest at the left upper sternal border, presence of an S3, and right-sided heave. EKG showed sinus tachycardia with right bundle branch block and QRS duration of 192 ms. Chest x-ray showed right ventricular enlargement and prominent pulmonary arteries.

Transthoracic echocardiogram revealed moderate right ventricular systolic dysfunction with severe dilation, severe right atrial enlargement, and septal flattening in diastole suggestive of RV volume overload. An overriding aorta as well as VSD patch repair without residual shunt were noted. Severe PR with branch pulmonary artery flow reversal was also seen. Cardiac MRI confirmed the presence of a valved-conduit with severe PR. The RV end diastolic volume index was calculated to be 200 mL/sq m, and the RV ejection fraction was estimated at 35%. The pulmonary annulus was measured at 35 mm.

Given the patient's progressive symptoms, he met criteria for intervention. Pulmonary valve replacement (PVR) via intra-cardiac surgery was chosen rather than trans-catheter approach due to the pulmonary annulus size. The patient underwent PVR with a 27mm Mosaic bioprosthetic valve. The surgery was complicated by a run of ventricular fibrillation lasting <30 seconds after opening the chest, which required compressions and defibrillation twice. The surgery was otherwise without complications, and his symptoms resolved during follow up.

Discussion

Pulmonary regurgitation is a late complication of Tetralogy of Fallot repair and has been linked to important adverse outcomes.[1] Chronic PR increases right ventricular preload and contributes to the remodeling of the right ventricle, leading to right ventricular dilation.[2] Right ventricular dilation has been directly linked to right heart failure and poor exercise tolerance, as experienced by this patient.[3] Right ventricular dilation has also been linked to significant arrhythmias and sudden death,[4] reflected in this patient's frequent palpitations. Arrhythmias in post-TOF repair patients are often predicted by absolute QRS prolongation to >180ms.[5] In our patient, his QRS duration was 192ms with morphology diagnostic of right bundle branch block.

Diagnosis of PR is made by 2-D and color Doppler echocardiogram with cardiac MRI being the gold standard in evaluation of PR. However, cardiac MRI is relatively expensive and not universally available. Therefore, standardized grading of PR severity using echocardiogram is becoming increasingly important in determining the timing of

pulmonary valve replacement. Diastolic flow reversal in main or branch arteries, PR jet width >50% of pulmonary annulus, PR pressure half time of <100 ms, and PR index of <0.77 are significant echocardiographic findings of severe PR.[6] Color Doppler assessment alone is insufficient as wide-open, low-velocity PR can be easily missed. In our patient, branch pulmonary artery flow reversal by color Doppler, in addition to a broad PR color jet, were present.

Pulmonary valve replacement is indicated in PR patients who have symptomatic sustained ventricular tachycardia or overt symptoms of exercise intolerance or right heart failure. Given our patient's arrhythmia and new onset symptoms of SOB, palpitations, and syncope, surgical valve replacement was performed. Pulmonary valve replacement has been shown to reduce right ventricular size, stabilize QRS duration, and reduce the incidence of subsequent ventricular tachycardia.[7]

Conclusion

As primary surgical repair of Tetralogy of Fallot becomes more successful, late complications such as pulmonary regurgitation become increasingly common. TOF patients require careful long-term follow-up in order to detect sequelae of surgical intervention early. Significant PR should be suspected in patients with signs or symptoms of right heart failure or arrhythmia and careful evaluation with a transthoracic echocardiogram should be the initial test of choice.

Figures

Figure 1: Transthoracic echocardiogram, color Doppler in parasternal short axis view in diastole. Severe pulmonary regurgitation with evidence of main and branch pulmonary artery flow reversal. RVOT = Right Ventricular Outflow Tract, Ao = Aorta, PR = Pulmonary Regurgitation, Main PA = Main Pulmonary Artery, RPA = Right Pulmonary Artery, and LPA = Left Pulmonary Artery.

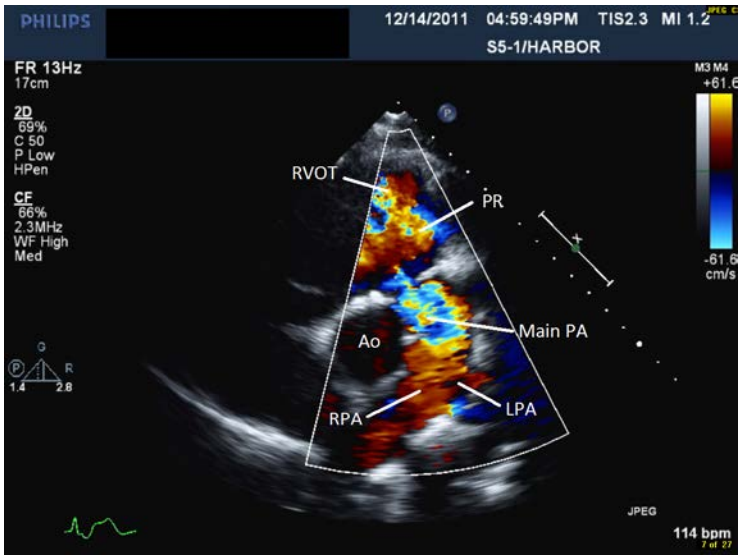
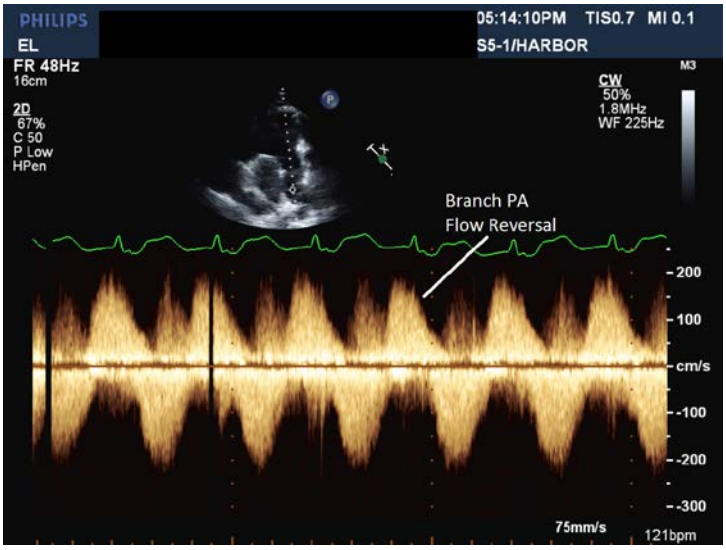


Figure 2: Transthoracic echocardiogram, continuous wave Doppler with sample volume in the LPA. Diastolic branch pulmonary artery flow reversal is seen. PA = Pulmonary Artery.



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