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Uncorrected Tetralogy of Fallot, Biventricular Dysfunction, and a Large Pericardial Effusion

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TETRALOGY OF FALLOT (TOF) is the most common cyanotic congenital heart disease, occurring in 1 of every 3,600 live births.[1] Patients with TOF commonly are diagnosed prenatally or early in life, with complete surgical repair performed by 6 months of life. Without surgical correction, 66% of patients with TOF live to 1 year of age, 49% to 3 years, 24% to 10 years, and 4% to 30 years.[2]

CASE REPORT

A 36-year-old male immigrant from Mexico, who had received limited medical care, presented with a several month history of progressive dyspnea, pedal and scrotal edema, orthopnea, and paroxysmal nocturnal dyspnea. His symptoms had worsened acutely during the prior 3 days, prompting him to seek medical attention. Transthoracic echocardiogram demonstrated cardiac anatomy consistent with TOF (Fig 1). The left ventricle showed global hypokinesis with an ejection fraction of 35%, a large ventricular septal defect, a hypokinetic and severely enlarged right ventricle (RV), subvalvular muscular right ventricular outflow tract (RVOT) obstruction with a peak gradient of 81 mmHg (Fig 2), and severe biatrial enlargement (right atrium measured 119 cm^2). A large pericardial effusion also was present (Fig 3). Electrocardiogram showed junctional bradycardia with a rate of 20 to 30 beats per minute when sleeping and 40 to 50 beats per minute when awake. Right-heart catheterization demonstrated elevated systemic RV pressures (117/24 mmHg) and a mean pulmonary artery pressure of 42 mmHg.

E-CHALLENGE

What is the best therapeutic management for an adult with uncorrected TOF and biventricular dysfunction?

What is the appropriate timing of surgical intervention in a patient with acute or chronic congestive heart failure?

The patient's case was discussed in a multidisciplinary meeting. The team decided to proceed with surgical correction of the patient's native anatomy. Insertion of a ventricular assist device and heart transplantation were precluded by the patient's lack of medical insurance.

An induction plan for a patient with TOF, biventricular dysfunction, and a large pericardial effusion was described. The optimal "rescue" cardiovascular medications and interventions for a patient with TOF and biventricular dysfunction were reviewed.

On presentation to the operating room, large-bore intravenous and radial arterial access were obtained. External pacing pads were placed in the event of worsening bradycardia while the patient was under anesthesia. General anesthesia was induced using midazolam, fentanyl, ketamine, and rocuronium. Volume-controlled ventilation was used, with tidal volumes of 350 to 400 mL and a peak inspiratory pressure of 46 cm H_2O. The capnogram indicated an expiratory obstructed ventilation pattern; the authors determined that severe cardiomegaly, with significant contribution from both atria, was the cause (Figs 4 and 5).

The patient's hemodynamics remained stable before cardiopulmonary bypass (CPB) was initiated. Because the patient's baseline hemoglobin was 16.3 g/dL, 2 units of whole blood were removed before heparin was administered.

After CPB was initiated, the patient underwent muscular resection of the RVOT, annuloplasty of the tricuspid valve, resection of a large strip from the right atrium (RA) free wall, and patch repair of the ventricular septal defect. The patient was weaned from CPB with epinephrine, 0.1 μg/kg/min, and milrinone, 0.25 μg/kg/min. Intraoperative transesophageal echocardiogram revealed mild tricuspid regurgitation, resolution of subvalvular stenosis, and moderately decreased biventricular function, and reduced RA dimensions (Fig 6). After RA resection, ventilation improved, with a reduction in peak inspiratory pressure from 46 to 26 cmH_2O and normal capnography findings.

Hemostasis was challenging in the postbypass period because of surgical bleeding at the RA suture line. The patient received autologous whole blood, 2 units of platelets, 3 units of fresh frozen plasma, 1 unit of packed red blood cells, and 3 mg of activated factor VII (administered in 1-mg doses). After hemostasis was achieved, the patient's chest was closed and he was transported, intubated and sedated as planned, to the cardiac surgical intensive care unit. Significant chest tube output in the first 12 postoperative hours necessitated transfusion of 5 units of allogeneic red blood cells, 3 units of cryoprecipitate, and 1 mg of activated factor VII. The patient did not require surgical re-exploration.

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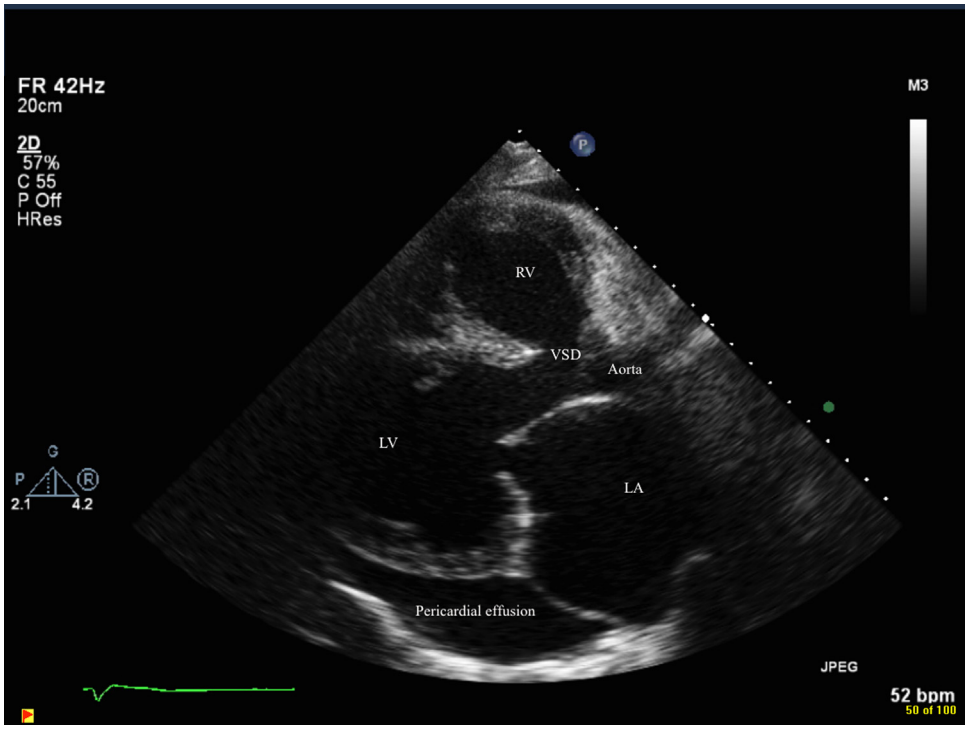

Fig 1. Preoperative transthoracic parasternal long-axis view demonstrating the VSD and overriding aorta in tetralogy of Fallot. LA, left atrium; LV, left ventricle; RV, right ventricle; VSD, ventricular septal defect.

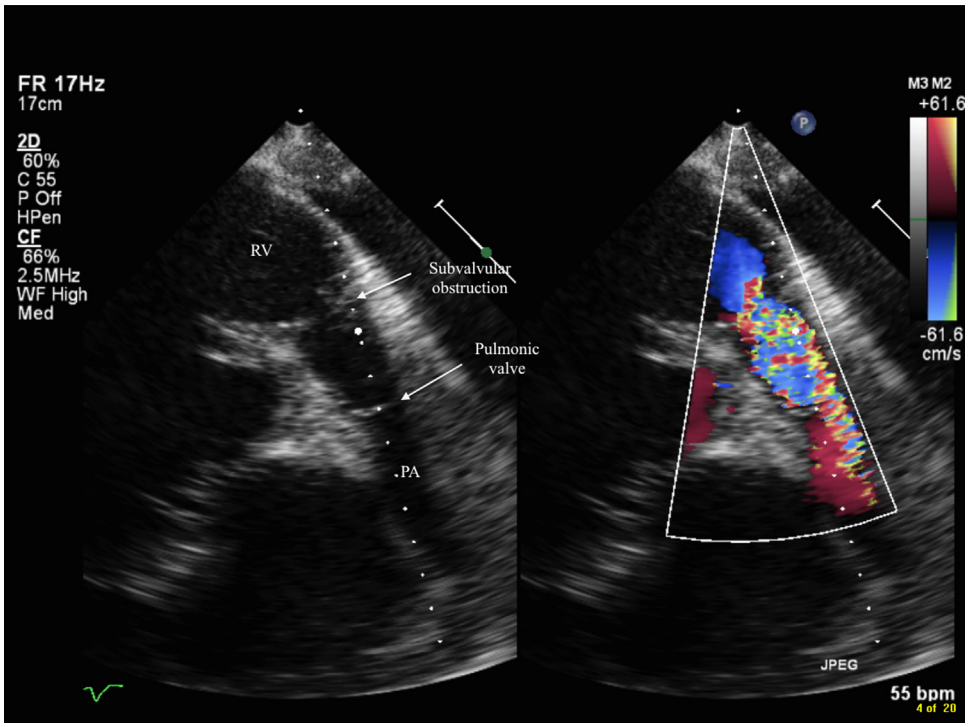

Fig 2. Preoperative transthoracic parasternal long-axis RV outflow view with 2D (left) and color-flow Doppler (right) demonstrating the subvalvular obstruction in the RVOT. PA, pulmonary artery; RV, right ventricle; RVOT, right ventricular outflow tract.