E-CHALLENGES & CLINICAL DECISIONS

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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 cm²). 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 H₂O. 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 cmH₂O 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.

Fig 3. Preoperative transthoracic parasternal short-axis basal view demonstrating a large pericardial effusion. MV, mitral valve; TV, tricuspid valve.

On postoperative day (POD) 1, the patient's airway was extubated and he was ambulatory, but he continued to receive inotropic support until POD 5. A permanent pacemaker-automatic internal cardiac defibrillator was placed on POD 4 because of pacer dependence and decreased left ventricular function. A postoperative chest radiograph showed significant reduction in cardiomegaly (Fig 7).

DISCUSSION

Although rare, cases of longevity in patients with uncorrected TOF are reported, with some patients living to >60

Fig 4. Chest radiograph demonstrating severe cardiomegaly, encompassing a large proportion of the cardiothoracic ratio.

years old.³⁻¹² Such patients often have aortopulmonary shunts or a small gradient across the RVOT obstruction, providing adequate pulmonary circulation and minimal symptoms. In published reports, only 1 patient presented with TOF and biventricular dysfunction in the setting of acute myocarditis; this patient achieved normal biventricular function with medical management alone.⁹

The authors present the first report of an adult with uncorrected TOF, severe biventricular dysfunction, and a large pericardial effusion who presented for surgical correction. Surgical and anesthetic management presented multiple challenges. First, the 30-day mortality in adult surgical correction of TOF is as high as 6%;¹³ though in the setting of acute or chronic heart failure and biventricular dysfunction, the risk of TOF is unknown but likely is increased. The nature and timing of surgical intervention were debated at length at a multidisciplinary conference. The procedure was preceded by 5 days of diuresis, permitting oxygen wean, and the administration of low-molecular-weight heparin as a result of the presence of spontaneous echo contrast shown on transthoracic echocardiogram.

Second, anesthetic management addressed conflicting hemodynamic goals of treating unrepaired TOF versus treating biventricular dysfunction. Induction agents included ketamine, midazolam, and fentanyl to maintain afterload. Because of the absence of tamponade physiology, positive-pressure ventilation was initiated despite the large pericardial effusion. The risk of ketamine-induced tachycardia in unrepaired TOF was negligible because of junctional bradycardia. Much thought was given to the choice of rescue medications in the event of hemodynamic instability. Goals in unrepaired TOF include maintenance of afterload, prevention of hypovolemia and

Fig 5. Transesophageal echocardiography midesophageal 4-chamber view demonstrating left and right atrial enlargement. LA left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Fig 6. Transesophageal echocardiography midesophageal 4-chamber view after the repair demonstrating smaller right atrial dimensions, closer to normal anatomy. LA left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Fig 7. Chest radiograph demonstrating significant reduction in size of the right cardiothoracic border compared with the preoperative chest radiograph.

tachycardia, and β -antagonism to relieve dynamic RVOT obstruction. Goals in biventricular dysfunction include β -agonism, afterload reduction, and prevention of hypervolemia to enhance ventricular ejection. Because of this patient's congestive heart failure, the authors prioritized ventricular

function and planned for epinephrine administration to treat hemodynamic instability.

Third, the patient's ventilatory disturbances presented interesting introspection. The extreme cardiomegaly created an obstructive respiratory pattern that was not identified preoperatively. Recognition of the obstructed ventilation pattern altered the surgical plan to include right atrial resection, which improved ventilation after CPB.

Finally, post-CPB hemostasis was challenging. Coagulation abnormalities in cyanotic CHD include thrombocytopenia, platelet dysfunction, factor deficiencies, fibrinogen dysfunction, and impaired fibrinolysis, inversely correlated to the degree of polycythemia.^{14,15} A form of disseminated intravascular coagulation has been proposed as a mechanism for such hemostatic disturbances. Increased viscosity from polycythemia results in vascular stasis and intravascular deposition of fibrin and platelet thrombi, leading to consumption of platelets and coagulation factors.¹⁴ Such disturbances likely contributed to excessive intraoperative and postoperative bleeding.

Unrepaired adult CHD presents new challenges to the surgical and anesthetic teams. Compensatory changes to the heart may fail, leading to acute heart failure. The resulting physiology may present conflicting hemodynamic goals and challenging ventilatory strategies, as highlighted by this patient's case. Thorough preparation and readiness to adapt intraoperatively are essential. Recognition of hemostatic concerns in cyanotic congenital heart disease is crucial to ensure preparedness of the institutional blood bank.

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