Infective Endocarditis with Multiple Valvar Vegetation in Uncorrected Tetralogy Fallot: A Case Report of Rare Condition

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	Abstract
	Objective: To describe a case of an adult patient with uncorrected Tetralogy of Fallot (TOF) who suffered from Infective Endocarditis (IE) that involved the tricuspid and aortic valves.
pISSN: 2302-1381; eISSN: 2338-4506; http://doi.org/10.15850/ ijihs.v9n2.2165 IJIHS. 2021;9(2):79–83	Methods: On October 2019, a-twenty-year-old male with uncorrected TOF was admitted to the emergency department of Dr. Hasan Sadikin General Hospital with a chief complaint of fever. The echocardiography showed the presence of vegetation on the tricuspid valve and aortic valve. The patient was given empirical antibiotics therapy for IE treatment. The serial blood cultures did not yield any organism growth and the transthoracic echocardiography evaluation showed healed vegetation after 21 days of antibiotics treatment.
	Results: A high turbulence in small VSD and valvular stenosis, as found in TOF, can injure the endocardial surface. While bacterial adherence to the injured endocardial surface can cause IE, no specific criteria available for diagnosing IE in congenital heart disease (CHD) patients compared to other population. In CHD patients, it is necessary to consider the diagnosis of IE presenting with signs of infection or fever. The patients was known to have uncorrected TOF and was admitted to the hospital with a chief
Received: October 12, 2020	complaint of fever. The uncorrected TOF was complicated by an unusual form of aggressive vegetation involving multiple valves.
Accepted: September 28, 2021	Conclusion: The incidence of IE in uncorrected TOF is around 4%. Patient is advised to receive a corrective surgery for the cardiac anomaly. Surgical repair of CHD with no residual lesion reduces the risk of IE.
	Keywords: Infective endocarditis, multiple valvar vegetation, tetralogy of fallot

Introduction

Infective endocarditis (IE) describes especially to an infection of the heart valves and endocardium or a prosthetic valvular implant. Infective endocarditis (IE) is a life-threatening disease, a rare and that has long-lasting effects even among patients who are cured and survived. Because of Tetralogy of Fallot (TOF)

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is associate multiple cardiac lesions, it carries a higher risk for IE. $^{\rm 1-3}$

The factors that influence the prognosis of IE: the cardiac and noncardiac complications, patient characteristics, the echocardiographic findings and the infecting organism. Based on 2015 ESC Guideline for the management of IE, there were three categories of patients with the highest risk of IE. First, patients that use prosthetic material or with a prosthetic valve² Second, patients with uncorrected CHD and CHD patients who have postoperative palliative shunts, prosthetic material or other conduits. Third, patients known with previous IE. These patients have a greater risk, higher incidence of complications and higher mortality and

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than patients with a new episode of IE.^{3,4} IE antibiotic prophylaxis is recommended for the first six months after surgical repair with no residual defects. The prophylaxis is given after the procedure until endothelialisation of the prosthetic material has occurred.² Presented here is an adult patient with uncorrected TOF that was complicated by an unusual form of aggressive vegetation involving multiple valves.

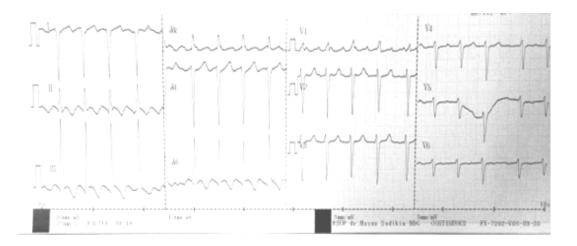
Case

On October 2019, a 20 years old man with known uncorrected TOF was admitted to the emergency department Hasan Sadikin Hospital with a chief complaint of fever for seven days before admission. He was aware of his congenital heart disease since he was twoyears-old and suggested surgical correction, but his parents refuse due to financial problem. There was a history of cyanosis on the lips and fingertips when activity and crying since a child. Growth and development are smaller compared to his peers at the same age. There is no history of frequent respiratory infections at the age of children. There was no history of family members with similar complaints. The patient had poor dental hygiene. He looked moderately ill, fully alert and dyspnea with obvious work of breathing. He was febrile, and his blood pressure was 110/60 mmHg, his pulse was 107 bpm with positive pulses deficit, and his respiratory rates was 26 bpm. The head examination showed perioral cyanosis. The jugular vein pressure showed 5+3 cm H20 with positive hepatojugular reflux. The

thoracal examination showed cardiomegaly with pansystolic murmur grade 3/6 at lower left sternal border with positive Carvallo's sign and systolic murmur grade 3/6 at the upper left sternal border. Other physical examinations were unremarkable. The electrocardiography examination showed atrial flutter, right axis deviation and biventricular hypertrophy (left ventricular hypertrophy with Sokolow lyon criteria, R/S ratio >1 and dominant S wave in V5 or V6). The laboratory examination showed Hb 18.3 g/dL, Ht 64.2%, leukocyte 12.040 / mm³, thrombocyte 165.000/mm³, ureum 88.8 mg/dL, creatinine 1.28 mg/dL. Chest X-Ray examination showed cardiomegaly without lung oedema and right loculated pleural effusion.

The patient underwent transthoracic echocardiography examination with result Situs solitus, AV-VA concordance. All PV drain to LA. ASD (-), PDA (-). VSD perimembranous (33-34 mm) R to L shunt with overriding aorta 50%. Dilated all chambers. Reduced LV systolic function with global hypokinetic. Diastolic dysfunction cannot be measured due to fusion and reduced RV contractility. Mild AR, trivial MR, mild TR. Severe PS valvar, PS gradient 88 mmHg. PA confluence, MPA diameter 14 mm, RPA diameter 9 mm, LPA diameter 8 mm. Left Ao arch, CoA(-). Mild pericardial effusion is circumferential with conclusion Tetralogy of Fallot and Vegetation at tricuspid valve (8x11 mm) and NCC of the aortic valve (6x12 mm).

The serial blood cultures did not yield any organism growth. Patients then diagnosed with *Tetralogy of Fallot, AFNVR, Acute Decompensated Heart Failure*, possible *IE* (Duke





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Fig. 2 Chest X-Ray Showed Cardiomegaly Without Lung Oedema and Right Loculated Pleural Effusion

criteria: 1 major criterion: Echocardiography positive for IE (vegetation), two minor criteria: the presence of cardiac abnormalities predisposing to IE, fever), and Right Pleural *Effusion* and were given Ampicillin sulbactam 4 x 3 gr iv, Gentamicin 1 x 160 mg iv, sildenafil 3 x 25 mg PO, methylprednisolone 1 x 19.2 mg iv, sucralfate 4 x 10 cc, omeprazole 2 x 40 mg iv. for treatment. He was symptom-free and went home after 21 days of hospitalisation. The echocardiography evaluation after 1 month showed healed vegetation (2x6 mm at NCC) in response to antibiotics treatment. He was advised periodic follow-up and advised corrective surgery for the cardiac anomaly.

Discussion

Congenital heart disease (CHD) is responsible for 10%–20% of IE cases, with tetralogy of Fallot and other uncorrected cyanotic congenital heart diseases.^{5,6} The incidence of IE in CHD patients has been increased.⁷ In spite of the improvement of antibiotics, IE is still high frequency, observed in the adult with CHD. It could be due to improvement in catheter intervention and cardiac surgery that increase in the survival rate of adult patients with complex CHD. The incidence of IE in corrected TOF is around 18%. The incidence of IE in uncorrected TOF is around 4%.⁵

As patients with TFF are born with a high risk of IE because of the nature of their CHD the exact pathophysiological mechanism remains speculative it is paramount to examine the risk of IE from the day of birth. In this case, the uncorrected TOF was complicated by an unusual form of aggressive vegetation involving multiple valves. Each CHD lesion has varying pathogenesis which leads to endocardial injury. Whereas, high turbulence in small VSD and valvular stenosis as in TOF can injure endocardial surface.⁵ The vegetations at tricuspid valve due to endocardial injury because high turbulence from VSD. The vegetations at aortic valve due to endocardial injury because overriding aorta. The pathogenesis of IE requires several independent factors such as injury of endocardial surface, thrombus formation, bacterial entry and adherence to the injured endocardial area. The injury and thrombus formation conditions provide а good environment to infection, whereas the next two allow the organism to implant on the endocardial area. The preexisting valvular

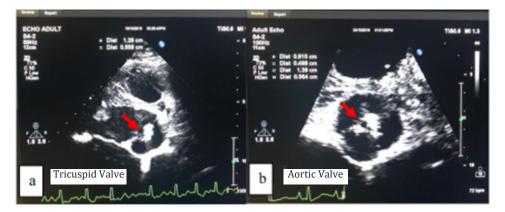


Fig. 3 Echocardiography Showed Vegetation at Tricuspid Valve in RV Inflow View (a), Vegetation at NCC of Aortic Valve in Short Axis View (b)

disease makes the turbulence blood flow. The turbulence makes injury of the endothelium. Mostly endocarditis patients have hemodynamic or structural abnormalities.⁸ While the endocardial surface is injured, platelets adhere to initiate sterile vegetation (called *nonbacterial thrombotic endocarditis*) formation from fibrin deposition in the injured endothelial surface.⁸

Nowadays, IE is usually an acute disease, that associated with healthcare, often with early presentations due to *Streptococcus aureus* in developed countries. Fever is nonspecific but is the most common presenting symptom nowadays.^{2,3} Patient's was admitted to the hospital with a chief complaint of fever. The serial blood cultures did not yield any organism growth. The patient was diagnosed possible *IE* (Duke criteria: 1 major criteria: Echocardiography positive for IE (vegetation), two minor criteria: the presence of cardiac abnormalities predisposing to IE, fever).

Right-sided infective endocarditis is more frequent in congenital heart disease than in acquired cardiac disease. However, the basic principal symptoms, diagnosis and complication same from IE in general.² The use of transesophageal echocardiography is favourable in an adult with artificial material and complex cardiac anatomy. The superiority of transesophageal echocardiography over transthoracic echocardiography has not been systematically studied in this setting. The antibiotic treatment is started after taking the blood culture.⁹The patient was given empirical antibiotics therapy for IE treatment. The serial blood cultures did not yield any organism growth. The transthoracic echocardiography

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evaluation showed healed vegetation after 21 days of antibiotics treatment.

In CHD, patients with IE should see CHD centres. The specialist CHD centres provide expertise in microbial examination, imaging, surgery and intensive care unit.² The principles of diagnostic and management in IE patients with or without CHD is same. The management is mainly appropriate antimicrobial treatment based on the respective microorganism.² Surgery is limited to those with failure of medical treatment, serious haemodynamic complications, IE with a prosthetic material, and when there is a high risk of devastating septic embolism.⁴ In another study, cardiac surgery should be considered in the presence of heart failure, a perivalvular abscess, aortic valve IE, and the change of antibiotics.¹⁰ The patient was advised periodic follow-up and advised corrective surgery for the cardiac anomaly.

Primary prevention for IE with CHD is vital. It is important to maintain good oral and dental hygiene and skin hygiene. In patients not followed in specialist, CHD centres could cause education problem about prevention of recurrent IE.¹¹ The patients are suggested to avoid piercing and cosmetic tattooing.

The limitation of this case report is we could not do pathological examination of resected valvar or embolic fragment because surgery was not performed as gold standard for diagnoses of IE. The incidence of IE in uncorrected TOF is around 4%. The patient was advised corrective surgery for the cardiac anomaly. Surgical repair of CHD with no residual lesion reduces the risk of IE.

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