A CHILD WITH TETRALOGY OF FALLOT PRESENTING WITH COMPLICATIONS OF INFECTIVE ENDOCARDITIS, CEREBRAL ABSCESS, AND UNDERNUTRITION

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ABSTRACT

Tetralogy of Fallot (TOF) is a congenital heart disease that consists of four anatomical anomalies, namely ventricular septal defect, pulmonary stenosis or obstruction of the right ventricular outflow tract, right ventricular hypertrophy, and overriding aorta. Tetralogy of Fallot is frequently associated with complications that could affect morbidity and mortality because of its complex cardiac lesions. Good understanding of the natural history and complications of this disease is very important to guide the management of patients. We reported a 4-year old girl with classic TOF with complications of infective endocarditis, cerebral abscess, and undernutrition. Despite optimal medical therapy, the patient's condition showed no satisfying improvement. Since the parents were refused to take more aggressive measure by undergoing surgical therapy, so that patient care remains focused on supportive and palliative aspects. **[MEDICINA 2015;46:37-41].**

***Keywords*:** *tetralogy of Fallot, infective endocarditis, brain abscess, undernutrition.*

SEORANG PENDERITA *TETRALOGY OF FALLOT* DENGAN KOMPLIKASI ENDOKARDITIS INFEKTIF, ABSES SEREBRAL, DAN GIZI KURANG

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ABSTRAK

Tetralogy of Fallot (TOF) merupakan suatu penyakit jantung kongenital yang terdiri dari empat anomali anatomis, antara lain *ventricular septal defect*, *pulmonary stenosis* atau obstruksi pada *right ventricle outflow tract*, hipertrofi ventrikel kanan, dan *overriding aorta*. *Tetralogy of Fallot* sering disertai dengan berbagai komplikasi yang dapat mempengaruhi morbiditas dan mortalitas karena kelainan ini merupakan kelainan kongenital kardiak yang kompleks. Pemahaman yang baik mengenai perjalanan alamiah dan komplikasi penyakit ini sangat penting untuk mengetahui penatalaksanaan pasien. Kami melaporkan seorang anak perempuan berusia 4 tahun dengan TOF klasik yang juga menderita komplikasi endocarditis infektif, abses serebral, dan gizi kurang. Walaupun telah memperoleh terapi medikamentosa yang optimal, kondisi pasien tidak menunjukkan perbaikan yang memuaskan. Orangtua pasien telah menolak pemberian tindakan yang lebih agresif, yaitu dengan terapi pembedahan, oleh karena itu penatalaksanaan pasien lebih difokuskan pada aspek suportif dan paliatif. **[MEDICINA 2015;46:37-41].**

***Kata kunci*:** *tetralogy of Fallot, endokarditis infektif, abses serebral, gizi kurang.*

INTRODUCTION

Tetralogy of Fallot (TOF) is a congenital heart disease that was first described by Etienne-Louis Arthur Fallot. In TOF, there are four anatomical anomalies, i.e ventricular septal defect (VSD), pulmonary stenosis (PS) or obstruction of the right ventricular outflow tract (RVOT), right ventricular hypertrophy, and overriding aorta.[1,2]

Tetralogy of Fallot is cyanotic congenital heart disease, which is most often found in both children and adults. The prevalence of TOF is 4-10% of all congenital heart disease.[3] Children with TOF bear increased risk for infective endocarditis. Infective endocarditis is a serious and fatal complications in patients with congenital heart disease.[1,4] Although there have been a lot of advances in the prevention and diagnosis of infective endocarditis as well as medical treatment and surgery for its treatment, the incidence of complications and mortality due to infective endocarditis is still high. The mortality rate of patients suffering from congenital heart disease was 8.8%, if complicated with infective endocarditis. One of the other complications that frequently occur in patients with TOF is a cerebral abscess.[5]

Cerebral abscess is an infection that occurs in the brain parenchyma. There were increased risk for the occurrence of

cerebral abscess in patients with cyanotic congenital heart disease due to hypoxic conditions which cause polycythemia and hyperviscosity. The presence of congenital heart disease with various complications could also cause problems in the intake and utilization of nutrients, so often happens undernutrition or malnourished condition. Along with medical and surgical treatment of these patients, developing a nutritional strategy plays a key role in the patient management.[6]

We presented a case report of TOF patient with complications of infective endocarditis, cerebral abscess, and undernutrition.

CASE ILLUSTRATION

A 4 year-old girl was referred to our hospital with suspected diagnosis of congenital heart disease. The patient complained of shortness of breath that were accompanied by bluish lips and fingertips since the age of 2. Shortness of breath with bluish arise while patients were too tired after playing. Shortness of breath was said to be improved if she was squatted. She also complained of fever since 21 days ago (at the time of previous hospitalization).

On the physical examination, the patient looked severely ill but still fully alert. The pulse was 118 beats / minute, axillary temperature was 39°C and respiratory rate was 40 breaths / min, oxygen saturation was 70-79% at nasal oxygen supplementation 1-2 liters per minute, weight was 40 kg, height was 100 cm (BMI: 22.56 kg / m^2). There was an increased JVP PR + 4 cm H2O. Heart examination obtained apex beat could be seen at ICS IV left midclavicular line, first and second heart sounds regular, palpable thrill in ULSB, systolic ejection murmur in ULSB, grade III / VI, with RV heave (+). There were cyanosis and clubbing fingers of all four limbs. The others was unremarkable. Laboratory examination revealed WBC 13.82 X 10^3 / mL, hemoglobin 15.3 g / dl, HCT 57.1%, MCV 67.38 fL, platelets 100 x 103 / mL, CRP 133 mg / L, sodium 135 mmol / L, potassium 4.10 mmol / L. Electrocardiography examination was concluded as sinus rhythm 100 times per minute with right axis deviation, right atrial and ventricular hypertrophy. The conclusion of chest x-ray were boot-shaped heart, concave pulmonary segment, with rounded apex.

The patient underwent echocardiographic examination with results of classic TOF, left aortic arch, major aortopulmonary collateral artery (MAPCA), patent foramen ovale (PFO), mild valvular regurgitation and aortic tricuspid, and there was vegetation resided between the right coronary cusp (RCC) of the aortic valve and the interventricular septum measuring 1.1 x 1.0 mm (**Figure 1**). She was diagnosed with TOF, infective endocarditis, and undernutrition, and acquired propanolol 13 mg TID, paracetamol syrup 1,5 tsp TID, and antibiotic therapy including gentamicin 8 mg / kg / day intravenously on the first day followed by gentamicin 6 mg / kg / day intravenously and ceftriaxone 100 mg / kg / day intravenously.

Patient was also consulted to the Division of Nutrition because of undernutrition and decreased food intake. Nutrition therapy provided were pediasure 3 x 200 cc and porridge diet 3 x 1 portion. On the 8th day of admission, the patient complained of fever, decreased of consciousness, appeared sleepy, and further decreased of food intake. Physical examination found somnolence alertness and the presence of clonus on both legs. She was suspected to suffer from cerebral abscess. Head CT scan was proceeded and obtained multiple hypodense lesions with firm border with varying sizes in the right and left parietal lobe of the brain parenchyma, could be an image of the infection process (**Figure 2**). She received additional treatment from Neurology

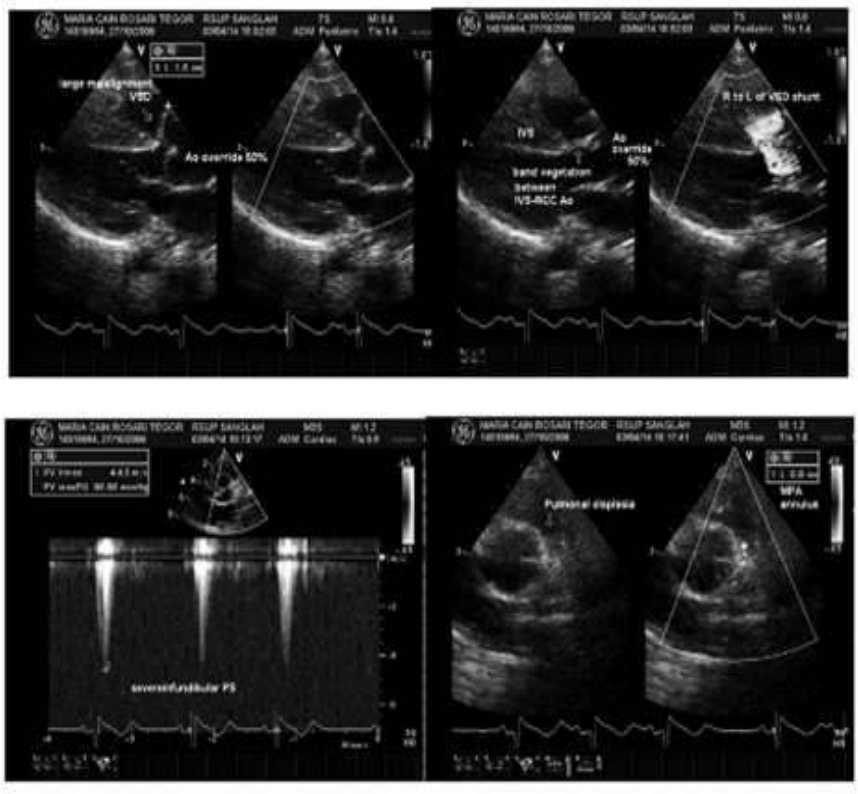

Figure 1. The echocardiography results of the patient.

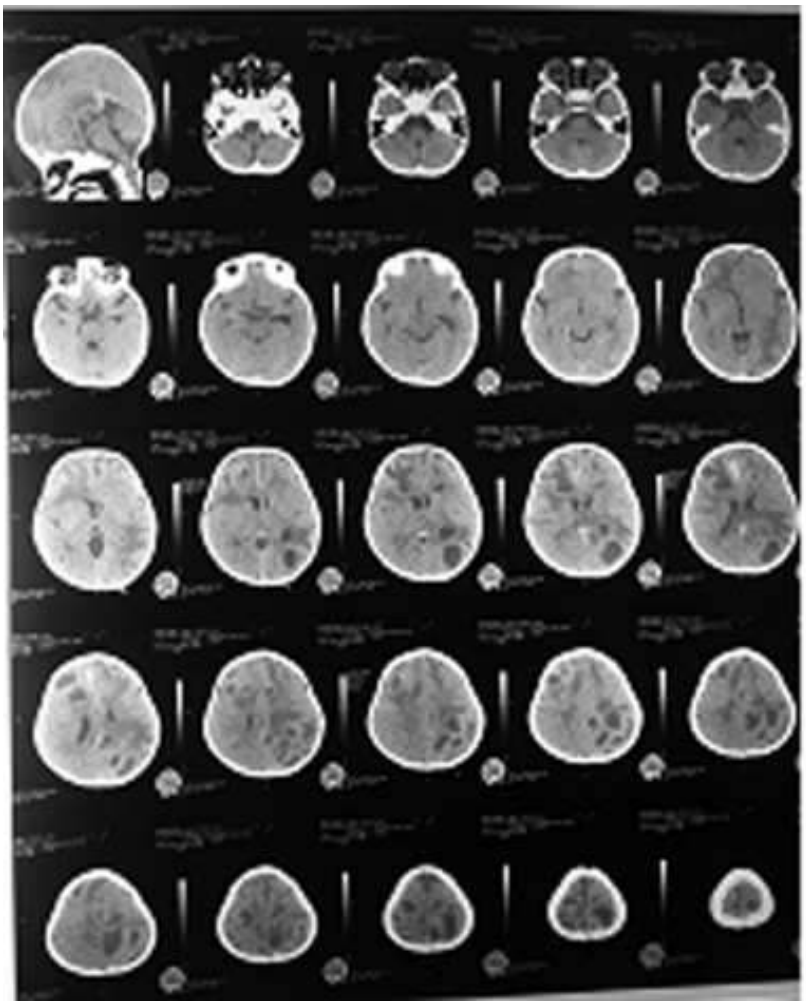

Figure 2. The head CT-scan result of the patient.

Department, i.e metronidazole loading of 15 mg / kg followed 7.5 mg / kg every 8 hours, chloramphenicol 100 mg / kg / day, and cefotaxime 200 mg / kg / day with dexamethasone loading of 1 mg / kg followed by 0.5 mg / kg / day and was advised to consult to Neurosurgery Department. Neurosurgery Department planned for burrhole and abscess drainage, but the parents were refused. Therefore it was decided to provide conservative and supportive management with continuing the antibiotics.

DISCUSSION

Tetralogy of Fallot is the most commonly found cyanotic congenital heart disease. Its incidence is estimated 10% of all congenital heart disease. Defects in TOF caused by ventricular septal deviation towards anterochepalad resulting in four classic disorders. TOF is commonly associated by complications which could affect the morbidity and mortality of the patients, such as infective endocarditis, cerebral abscess, and undernutrition.[7,8]

Tetralogy of Fallot is a congenital heart disease with the highest risk of developing infective endocarditis because there are multiple cardiac lesions. There are two factors which play an important role in the pathogenesis of infective endocarditis, namely the area of endothelial damage and condition of bacteremia. One cause of bacteremia is dental hygiene and poor oral and dental caries, that was found in this patient.[9,10]

Clinical manifestations of infective endocarditis in children usually are not typical, and including longstanding fever and various somatic complaints. Although these complaints are nonspecific findings, the presence of these symptoms in certain circumstances such as in patients with congenital heart disease require careful evaluation for the presence of infective endocarditis. Modified Duke criteria are the most commonly used criteria for the diagnosis of infective endocarditis.[10,11]

Treatment of infective endocarditis in congenital heart disease is the same with the therapy in general. Indications of surgery are when there is a failure to medical therapy, if there is severe hemodynamic complications, and when there is a high risk of septic emboli. Primary prevention is very important for patients with congenital heart disease. Surgical correction of congenital heart disease may also reduce the risk of infective endocarditis if there is no residual lesions.[12,13]

Patient in this case report was diagnosed as definite infective endocarditis based on the modified Duke criteria for fulfilling one major criterion and 3 minor criteria. Although the patient did not show satisfying progress during the administration of antibiotic therapy, the parents already refused surgical treatment, so that patient care remains focused on supportive and palliative aspects.

Cerebral abscess is a serious infection of the brain parenchyma and could be life threatening. In normal conditions, the brain parenchyma is usually resistant to infection. The presence of focal ischemia or necrosis is a predisposing condition for the invasion by microorganism. It can be caused by episodes of hypoxia or embolization associated with cyanotic heart disease. Additionally, in TOF there was right-to-left, so that the venous blood does not pass through the

pulmonary circulation and can directly lead to bacteremia, septicemia, and the incidence of infective thromboemboli, especially in the brain. Cerebral abscess in patients with TOF often multiple and located at the border between gray matter and white matter. Abscesses are more often found in the parietal lobe because it has a large caliber and relate directly to the middle cerebral artery.[14]

Clinical manifestations which can occur in patients with cerebral abscess is a classic triad consisting of fever, headache, and focal neurological defects. The diagnosis can be confirmed by imaging examinations such as CT, MRI, and radioisotope brain scans. In cerebral abscess associated with cyanotic heart disease, first-line antibiotics is penicillin and cefotaxime, chloramphenicol, or metronidazole and / ceftriaxone therapy, with treatment duration between 4-6 weeks or depending on the organism involved and response to treatment. In patients also could be administered phenytoin, dexamethasone, or mannitol to reduce intracranial pressure. Indications surgery on cerebral abscess is an abscess size that does not shrink with antibiotics for 4 weeks, mass effect and significant neurological deficits, multiple lesions in locations covered by surgery, multiloculated lesions, lesions in the posterior fossa, and the diameter of the abscess is more than 2.5 cm.[14,15] Patients in this case report was suffered from cerebral abscess as a complication of TOF were diagnosed based on clinical manifestations of fever, in conjunction with the results of the CT-scan examination which revealed multiple abscesses at parietal lobe.

Undernutrition is a common complication in children with congenital heart disease, regardless of the type of defect and the presence or absence of cyanosis. In children with congenital heart disease usually suffered from loss of body mass that will exacerbate myocardial function and ventilation, healing capacity, and the immune system, resulting in increased risk of infection.[16,17]

The management of patients with congenital heart disease with undernutrition is very important to maintain adequate nutritional status. However, despite being given an aggressive nutrition programs, most children with congenital heart disease are not able to ingest enough calories to achieve or maintain a normal weight. In such cases, it is advisable to provide nutrients directly to the gastric using enteral nutrition. Continuous enteral nutrition can improve the intake of calories and nitrogen, preventing the occurrence of gastric distension and vomiting, increase the absorption of nutrients, and reducing the need for metabolism.[18,19] The patient in this case report is included in the criteria for undernutrition by Waterlow (83%) and obtained nutritional therapy in the form of 3 x 200 cc pediasure and porridge diet 3 x 1 portion. Nutritional therapy is switched to pediasure 150 cc every 4 hours via nasogastric tube, spent within one hour after the patient experienced a loss of consciousness due to cerebral abscess condition.

SUMMARY

We have reported a case of child who was suffered from classic TOF with complications of infective endocarditis, cerebral abscess, and undernutrition. The diagnosis of TOF, infective endocarditis, and undernutrition were made at the first time of admission by history taking and physical examination, along with echocardiography and laboratory examination. Despite optimal medical therapy, the condition of patient did not show satisfying progress, and she was also diagnosed with cerebral abscess on the 8th day of admission based on clinical manifestation and head CT-scan result. The parents already refused surgical treatment, therefore it was decided to provide conservative and supportive management with continuing the antibiotics.

REFERENCES

1. Sousa P, Santos W, Marques N, Cordeiro P, Ferrinha R, Pereira S, *et al*. A 72-year-old woman with an uncorrected Tetralogy of Fallot presenting with possible pulmonary endocarditis: a case report. J Med Case Rep. 2013;7(150):1-4.
2. Chandrasekaran B, Wilde P, McCrea WA. Tetralogy of Fallot in a 78-year-old man. N Engl J Med. 2007;357 (11):1160-1.
3. Atiq M, Ahmed US, Allana SS, Chishti KN. Brain abscess in children. Indian J Pediatr. 2006;73(5):401-4.
4. Pekdemir H, Cin VG, Akkus MN, Doven O. Cyanotic Tetralogy of Fallot with its infective endocarditis complication on the tricuspid and pulmonary valves of a 55-year-old man. Circ J. 2004; 68:178-80.
5. Niwa K, Nakazawa M, Tateno S, Yoshinaga M, Terai M. Infective endocarditis in congenital heart disease: Japanese national collaboration study. Heart. 2005; 91(6):795-800.
6. Mehnaz A, Syed AU, Saleem AS, Khalid CN. Clinical features and outcome of cerebral abscess in congenital heart disease. JAMC. 2006; 18(2):21-4.
7. Jacob G, Mathews C. Unrepaired Tetralogy of Fallot presenting with brain abscess. Calicut Med J. 2010;8(3):1-3.
8. Therrien J. Tetralogy of Fallot. In: Warnes CA, editor. Adult congenital heart

disease. USA: American Heart Association; 2009. p. 139-56.

9. Anwar AM, Nosir YF, Ajam A, Mushtaq M, Alama MN, Chamsi-Pasha H. Multivalvular infective endocarditis in a Tetralogy of Fallot. Echocardiogr J. 2008;25(1): 88-90.
10. Silva PV, Marinho JC, Martins P, Santos I, Pires A, Sousa G, *et al*. Infective endocarditis as a form of late presentation of congenital heart disease. Rev Port Cardiol. 2013;32(2):145-8.
11. Baddour LM, Wilson WR, Bayer AS, Fowler VG, Jr., Bolger AF, Levison ME, *et al*. Infective endocarditis: diagnosis, antimicrobial therapy, and management of complications: a statement for healthcare professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, and the Councils on Clinical Cardiology, Stroke, and Cardiovascular Surgery and Anesthesia, American Heart Association: endorsed by the Infectious Diseases Society of America. Circulation. 2005;111(23):394-434.
12. Harrison JL, Prendergast BD, Habib G. The European society of cardiology 2009 guidelines on the prevention, diagnosis, and treatment of infective endocarditis: key messages for clinical practice. Pol Arch Med Wewn. 2009;119(12):773-6.
13. Routray SS, Raut K, Mishra D, Mishra R. Cerebral abscess in a 8 years old with uncorrected Tetralogy of Fallot: Anaesthetic challenge. IJBAR. 2013;4(11):843-5.
14. Lakshmi V, Umabala P, Anuradha K, Padmaja K, Padmasree C, Rajesh A, *et al*. Microbiological spectrum of brain abscess at a tertiary care hospital in South India: 24-year data and review. Patholog Res Int. 2011;2011: 583139.
15. Ghafoor T, Amin MU. Multiple brain abscesses in a child with congenital cyanotic heart disease. JPMA. 2006;56(12):603-5.
16. Monteiro FP, de Araujo TL, Lopes MV, Chaves DB, Beltrao BA, Costa AG. Nutritional status of children with congenital heart disease. Rev Lat Am Enfermagem. 2012; 20(6):1024-32.
17. Sjarif DR, Anggriawan SL, Putra ST, Djer MM. Anthropometric profiles of children with congenital heart disease. Med J Indones. 2011; 20(1):40-5.
18. Rodica T. Nutritional approach of pediatric patients diagnosed with congenital heart disease. Acta Med Marisiensis. 2013;59(2):121-5.
19. Hubschman LE. Malnutrition in congenital heart disease: management to improve outcomes. ICAN. 2013;5:170-6.