A Rare Case of Embolic Stroke in Patient with Uncorrected Pentalogy of Fallot: A Case Report

Yusra Pintaningrum[1,2*] and Ketut Angga Aditya Putra Pramana[1]

[1]*Faculty of Medicine, Mataram University, Mataram, Indonesia*
[2]*Cardiology and Vascular Department of West Nusa Tenggara General Hospital, Mataram, Indonesia*
**Corresponding author. Email: yusra@unram.ac.id*

ABSTRACT
Pentalogy of Fallot is cyanotic type congenital heart disease that characterized with anatomical anomalies in Tetralogy of Fallot and atrial septum defect or patent foramen ovale. That condition makes cyanosis because of diminished oxygenated blood flow to the organs. Cerebrovascular accidents like embolic stroke are very rare among children. And can occur in patient with Pentalogy of Fallot because of thrombogenicity condition. In this article we describe a 2-year-old girl came to hospital with chief complaint suddenly left-sided paralysis. She had history of uncorrected Pentalogy of Fallot since she was born. Head CT scan with contrast were obtained to identified the affected brain area. She was given an antiplatelete and there is a slight improvement of paralysis was reported. Acute management to this condition can reduce the mortality and morbidity of the patient.

Keywords: *Pentalogy of Fallot, cerebrovascular accident, embolic stroke, antiplatelet*

1. INTRODUCTION

Pentalogy of Fallot which is cyanotic type congenital heart disease that characterized with anatomical anomalies in Tetralogy of Fallot and atrial septum defect or patent foramen ovale [1-3]. The first clinical sign of pentalogy of Fallot is cyanotic spells. This condition caused by shunting from right to the left of deoxygenated blood and blood flow obstruction of pulmonary artery which deteriorate after ductus arteriosus closed because of decreased pulmonary perfusion [2,4]. Episodes of cyanosis might be experienced by the patient during eating, defecating, crying and portrayed by serious cyanosis and often accompanied by tachypnoea. Impaired of consciousness and death might happen if the cyanotic episode is prolonged and more severe. Children may do a knee-chest position after physical activity as reduced of systemic vascular resistance and pressure of left ventricular while physical activity consequently increase the shunt from right to the left of blood so reduced of lung perfusion causing hypoxia and cyanosis. The stenosis degree at pulmonary or infundibular are related to a systolic murmur that can auscultate at the left upper sternal border [4].

Patient with untreated and serious disease might be complicate to impediment of growth or development and clubbing fingers. From laboratory assessment we can found erythrocytosis, relative iron insufficiency, and increment of erythropoietin because of hypoxia. [4] On radiography assessment we can found a boot-shaped heart that formed from hypertrophy of right ventricular makes upward of the cardiac apex, pulmonary conus collapse, and reduced of lung vascularity. Electrocardiography assessment show presence of right axis deviation and right ventricular hypertrophic. All anatomical abnormalities in pentalogy of Fallot can be assess by two-dimensional echocardiography examination [4].

Neurological complication in congenital heart disease contribute extensively to mortality and morbidity. Severe cerebrovascular accidents like stroke are the most common complication in many patients with uncorrected congenital heart defect [4]. Stroke cases happen in 25-30 percent of all patients with congenital heart disease with majority cases happen in

uncorrected disease [4,5]. In this case report article, we reported an uncommon case report of a 2-year-old child who had history of uncorrected Pentalogy of Fallot with complication of embolic stroke.

2. CASE REPORT

A 2-year-old girl child came to the hospital with complaints suddenly paralysis of her left-side extremities when she was playing according to her parents. This condition seems like similar in clinical signs of stroke. There is no any complaints of nausea and vomiting, loss of consciousness, or seizure. She had history of uncorrected cyanotic congenital heart disease Pentalogy of Fallot since she was born. Because of that condition, she always feel tired quickly when do some activities. She was routinely consuming 4 mg tablets propranolol three times in a day since diagnosed with that condition. There is no bad obstetric history during pregnancy according to her mother. From physical examination we obtained normal vital signs. On general physical examination we found cyanosis especially on her mouth mucosa and also, we found cyanosis with clubbing fingers in all her extremities. On heart sound auscultation we heard a systolic ejection murmur in the pulmonic area. On neurological motoric examination, the patient had power of +2 in both left-side extremities, her physiological reflex on left side extremities were increase and also, we found positive of Babinski pathological reflex. Reflex evaluation and motoric power on the right-side extremities were normal. From laboratory blood test examination, we obtained increase of haemoglobin, erythrocyte, leukocyte, and haematocrit (Table 1). Echocardiography examination to assess the heart anatomical abnormalities are showed in Figure 1, 2, and 3. Head computed tomography scan (CT scan) with contrast showed embolic infarction on right middle cerebral artery and no evidence of a haemorrhage (Figure 4).

Table 1. Basic blood test of the patient checked in West Nusa Tenggara General Hospital

Basic Blood Test			
Hb	16.7	Neu	37.9
RBC	$6.13x10^6$	Eos	1.7
WBC	18460	Baso	0.4
PLT	191000	Limf	41.8
Hct	49	Mono	18.2
BUN	11	SGOT	101
Scr	0.9	SGPT	23
GDA	77		

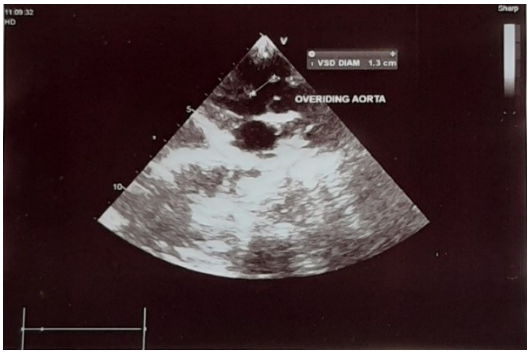

Figure 1. Defect of ventricular septum and overriding aorta on echocardiography examined in West Nusa Tenggara General Hospital.

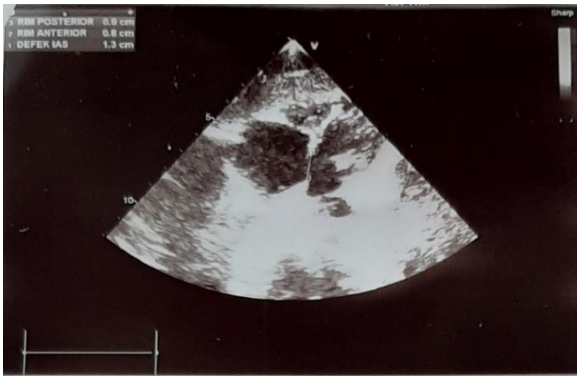

Figure 2. Defect of secundum atrial septum on echocardiography examined in West Nusa Tenggara General Hospital.

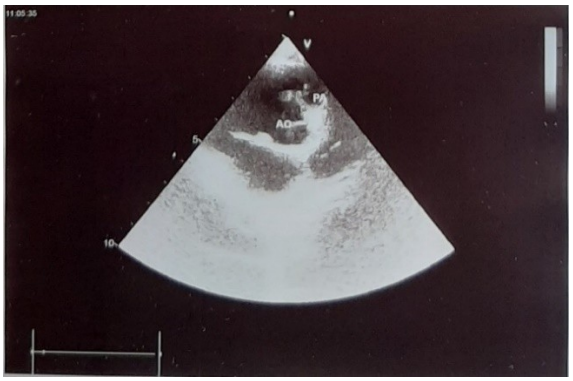

Figure 3. Severe Stenosis of pulmonic valve on echocardiography examined in West Nusa Tenggara General Hospital.

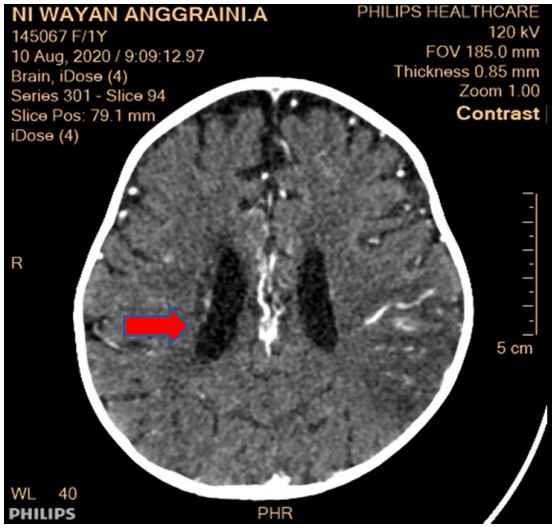

Figure 4. Head CT scan with contrast showed embolic infarction (arrow) examined in West Nusa Tenggara General Hospital

After we give the patient treatment with 4 times in one day 75 mg tablets acetyl salicylic acid, the patient had a slightly improvement of neurological manifestation and now she is able to move her left side extremities and had an improvement in her motor strength to +4 in the both of her left-side extremities.

3. DISCUSSION

Pentalogy of Fallot which is cyanotic type congenital heart disease that characterized with anatomical anomalies in Tetralogy of Fallot and atrial septum defect or patent foramen oval [1-3]. Diagnosis of Pentalogy of Fallot in this patient was based on patient's history and confirm with patient's clinical signs and the echocardiography examination [2,4]. Neurological complication in congenital heart disease contribute extensively to mortality and morbidity. Severe cerebrovascular accidents like stroke are the most common complication in many patients with uncorrected congenital heart defect.[4] Stroke cases happen in 25-30 percent of all patients with congenital heart disease with majority cases happen in uncorrected disease [4,5]. Diagnosis of embolic stroke in this patient were confirmed with head CT scan assessment. Paradoxical embolization and hyper viscosity in patient with uncorrected Pentalogy of Fallot are the most risk factor that can cause stroke with thromboembolism mechanism [6-8].

In paradoxical embolization, the emboli pass into the arterial circulation and systemic circulation without pass the vascular bed of lung circulation because of the right to the left shunting.[7,8] Hyper viscosity in cyanotic congenital heart disease caused by polycythaemia and compensatory erythrocytosis with haematocrits more than 70% which can increase the thrombogenicity events.[8] Persistent hypoxemia in cyanotic congenital heart disease can causing endothelial injury by initiate the activation of polymorphonuclear cells also releasing vasoactive and chemotactic factors. Then, platelets and endothelial cells collaborate and start the haemostasis process and activates the coagulation cascade [9].

We should consider clot prevention and correction of the shunt for management of paradoxical embolism, but correction of the shunt had no more benefit compared with administration of antiplatelet or anticoagulation to diminished the thrombogenicity events like stroke. Anticoagulation is proposed for congenital heart disease patients that have higher risk for embolic stroke. Low molecular weight heparin (LMWH) or warfarin is recommended for 1 year or until the cardiac anatomical abnormalities have been corrected. Aspirin should be considered for children suspected with cardiac embolism and lower risk for recurrent embolic stroke [7,8,10]. Phlebotomy should be considered for treatment of hyper viscosity, but it contraindication in congenital heart disease phlebotomy because can prompt iron deficiency and spherocytosis that can increase the risk the complication to stroke [8].

In this case report, acetyl salicylic acid tablets that administrated to the patient was not enough to improve neurological condition. After administration of that tablets, the patient only had slightly improvement of her neurological manifestation. After that, for reduce the morbidity and improve quality of life of this patient, we refer the patient to the medical rehabilitation clinic. This case was first case in our hospital with embolic stroke due to Pentalogy of Fallot.

4. CONCLUSION

Embolic stroke that caused by uncorrected pentalogy of Fallot is a complex and uncommon case. Embolic stroke in pentalogy of fallout can occur by allowing the paradoxical embolization. Early diagnosis and good intervention with anticoagulation or antiplatelet and total correction can treat and prevent worsening of neurological conditions and prevent the re-occurrence of embolic stroke.

ETHICAL APPROVAL

The study is in compliance with the Declaration of **Helsinki**.

CONSENT

The author **has** affirmed during submission that patient consent has been signed and collected in accordance with the journal's patient policy.

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