

# Hypercyanotic spell after general anesthesia in Tetralogy of Fallot

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## Abstract

A 21-year-old woman with tetralogy of Fallot referred because of a right temporo-occipital abscess, 2-months history of progressive visual loss, and headache. She had history of multiple drainages of brain abscesses and a Blalock-Taussing shunt, which was failed. With general anesthesia (GA) she underwent a temporo-occipital craniotomy and en-block removal of abscess. After termination of GA, she experienced two generalized seizures and became unconscious, tachycardia, and respiratory distress. The ABG showed low PaO<sub>2</sub> and low pH. The respiratory distress was controlled by propranolol. When hypercyanotic spell occurs during anesthesia or early postoperative period, could cause serious problems for diagnosis and treatment.

## INTRODUCTION

The hypercyanotic spell is common within the first 5 months of life in patients with tetralogy of Fallot (TOF). With increasing age, frequency of spell decreases and is rare in adulthood [1]. We present hypercyanotic spell in a 21-year-old TOF woman operated for brain abscess and its management.

## CASE REPORT

A 21-year-old TOF woman referred because of 2-month history of progressive visual loss and headache. On admission, the body weight was 35 kg. Her blood pressure was 90/60, pulse rate 110 / min, respiratory rate 21/min, and temperature 37°.

There was no jugular vein distention, hepatomegaly, or pedal edema. Lungs were clear. A thrill was palpable over left precardium. The neurological exam was normal, except for left homonymous hemianopsia and right lower quadrant visual field defect.

Her medical history was significant for multiple drainages of brain abscesses through burr holes with local anesthesia. A Blalock-Taussing shunt which was failed was carried out four years ago. She did not report any hypercyanotic spell during the last 10 years.

The lab results were summarized in table 1. Chest x-ray showed right ventricular hypertrophy. The echocardiogram

showed normal left atrial pressure, overriding of aorta, large bidirectional ventricular septal defect, hypoplastic pulmonary artery, normal left ventricular function with ejection fraction of 63%, and nonfunctioning Blalock-Taussing shunt.

## Figure 1

Table 1: Laboratory data before and after operation

Lab	Preoperative value	Early postoperative value
pH	7.29	7.023
PaCO <sub>2</sub> (mmHg)	34.2	58
HCO <sub>3</sub>	15.8	14.7
PaO <sub>2</sub> (mmHg)	45.7	31.4
SaO <sub>2</sub>	73.9%	55.7%
Hgb (g/dl)	65%	57%
Platelet (/μl)	210000	130000
Fasting blood sugar (mg/dl)	74	124
BUN (mg/dl)	13	7
Creatinine (mg/dl)	0.5	0.5
Sodium (meq/l)	145	135
Potassium (meq/l)	4.6	3.9
Calcium (mg/dl)	8.8	8.6

Brain CT scan showed a round large hypodense lesion with calcified enhancing perilesional ring in right temporo-occipital region. MRI demonstrated the round heterogenous lesion with a thick hypodense ring in T1WI and T2WI in

right temporo-occipital lobe.

With general anesthesia (GA) she underwent a temporo-occipital craniotomy and en-block removal of abscess which had a calcified thick wall with pus. Midazolam, ketamine, and fentanyl were used for GA induction and N<sub>2</sub>O, halothane, and fentanyl for GA maintenance. The GA and craniotomy was without problem. After operation she became awaked slowly in recovery room, extubated, and transferred to Neurosurgical ICU. In ICU she experienced two generalized seizures. Seizures were controlled with midazolam, phenytoin, and phenobarbital. After the seizures, she experienced loss of consciousness, tachycardia, and respiratory distress. The ABG showed low PaO<sub>2</sub> (31.4 mmHg) and pH (7.023). Serum biochemistry tests were normal (table 1). Brain CT scan showed total removal of abscess with small subarachnoid hemorrhage. The patient was on Ventury of 35% O<sub>2</sub>. Bicarbonate and morphine were administered. Respiratory distress was not responded to treatment until intravenous administration of propranolol, resulting in slowly resolving of the condition.

Within first week of operation she became conscious, and did not experience further seizure and any new neurological deficit. The respiratory and hemodynamic states were stable. After two weeks of antibiotic therapy she was discharged in good condition.

### DISCUSSION

Patients with TOF are at risk of different neurological complications, such as chronic cerebral hypoxia, impairment of neurological development, brain abscess, cerebrovascular thrombosis, and hemorrhage [1,2].

Anesthesia of patients with TOF and neurological complications is a demanding work. The general strategy, which is to avoid hypoxemia, is made by (1) ensuring adequate hydration, (2) maintaining systemic arterial blood pressure, (3) minimizing additional resistance to pulmonary blood flow, and (4) avoiding sudden increase in systemic oxygen demand (cry, inadequate level of anesthesia, seizure) [1]. This strategy would provide adequate perfusion to the hypertrophied right ventricle, prevent an increase in right-to-left shunting, and maintain collateral circulation into the lungs [3]. For these reasons, anesthesia of our patient was carried out with high-dose narcotic agents, benzodiazepines, and ketamine. Adequate hydration was achieved with normal saline and arterial blood pressure was maintained constantly with phenylephrine at the level of 90/60.

During the postoperative period, multiple seizures predisposed our patient to increase oxygen demand, decrease arterial PaO<sub>2</sub> and pH, and increase PaCO<sub>2</sub>. These factors were sufficient to lead our patient to hypoxemia and a hypercyanotic spell associated with hyperventilation, increasing venous return, increasing right-to-left shunt, and further decreasing arterial PaO<sub>2</sub> and pH. As postoperative neurological complications may have clinical manifestation similar to hypercyanotic spell, rule out of neurological problems is mandatory. Neuroimaging, proper physical examination, and laboratory results would be helpful.

Initial treatment of hypercyanotic spell should be aimed at reversal of right-to-left shunting [3]. Management and prevention of predisposing risk factors, like seizure, is helpful to block the vicious circuit of hypoxemia. Maintaining mean arterial blood pressure above 60 and restoring adequate circulating blood volume by infusion of intravenous crystalloid solutions are mainstays of treatment. Phenylephrine increases the mean arterial blood pressure and systemic vascular resistance, decreases the right-to-left shunt, and enhances pulmonary blood flow through aortic collateral vessels [1,3,4].  $\beta$ -blockers, particularly esmolol, are useful in reversing hypoxemic attack by decreasing right ventricle contractility and minimizing the infundibular obstruction [3,5,6]. Intravenous sodium bicarbonate has successfully terminated the spell with correcting peripheral metabolic acidosis and decreasing systemic vascular resistance [1]. Morphine and CPAP ventilator support also have been suggested for management [1,3].

### CORRESPONDENCE TO

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