

Perioperative Management of Undiagnosed Methaemoglobinaemia

Dr. Mahendrakar Sreevani¹, Dr. Rajapriya .R²

Abstract: *Methemoglobinemia is a rare and potentially life - threatening medical emergency that can be overlooked when evaluating a patient in respiratory distress. It is one of the causes for unexplained cyanosis with dark - coloured blood, especially in the absence of cardiac or pulmonary pathology. Without early recognition, patients may develop respiratory failure and die. This case report describes a child who posted for tympanoplasty who presented with 'chocolate - coloured blood' in the surgical field, despite blood gas analysis showing a normal partial pressure of oxygen.*

Keywords: Methemoglobinemia, 'chocolate - coloured blood, G - 6PD deficiency, desaturation, methylene blue

1. Introduction

There are many causes of hypoxia in the perioperative period and in the intensive care unit (ICU) ranging from airway obstruction to hemoglobinopathies. Methaemoglobinaemia, are rare but important cause of dark - coloured blood with or without clinically apparent cyanosis.

2. Case Report

A 11 - year - old child weighing 40 kg presented to the ENT Department with loss of hearing. Developmental milestones and intelligent quotient were normal. Cardiorespiratory system and airway examination were normal. Laboratory reports were within the normal limits. He was scheduled to undergo Tympanoplasty surgery. On the morning of surgery preop vitals HR 100 beats/min, BP 100/60 mmHg. Premedication - Glycopyrrolate IV (5 µg/kg), Fentanyl IV (2 µg/kg), Ondansetron IV (0.1 mg/kg). Induction - Propofol IV 2 mg/kg Atracurium IV 0.5 mg/kg, followed by intubation with a 6 - mm ID cuffed endotracheal tube. Initially, anesthesia was maintained with sevoflurane 2-3% in a mixture of 50: 50 oxygen and nitrous oxide Following incision, the surgeon noticed 'dark - coloured blood' in the surgical field, and a decrease in SpO₂ from 94% to 89% was noticed by the anaesthetist. Immediately, the FiO₂ was increased to 100% and the position of the endotracheal tube was checked. Good bilateral air entry and a good capnographic waveform confirmed optimum ventilation. An immediate arterial blood gas (ABG) sample was drawn that appeared 'dark coloured', but had the following results: PaO₂: 324 mmHg, PCO₂: 22 mmHg, HCO₃: 24 mmol/L and pH: 7.43. Then we suspected the patient might have abnormal haemoglobin, Methemoglobinemia was suspected because of the mismatch of SpO₂ and SaO₂, and surgery was post - poned for further examination. Cooximeter analysis revealed 21% methaemoglobin. Haemoglobin (Hb) electrophoresis and G6PD levels were normal. Erythrocyte nicotinamide adenine dinucleotide (NADH) - dependent methaemoglobin reductase level was low at 20.5 IU/g Hb (normal 35 ± 5 IU/gm Hb). A history of consanguinity was revealed on further questioning. He was promptly started on I. V. methylene blue (2mg/kg i. v. for 5 - 10 min) and oral ascorbic acid (200 mg/day) for 1 week. Surgery was rescheduled following 1week of methylene blue therapy,

after which, his methaemoglobin levels decreased to 1.2% and pulse oximetry showed 100% saturation. Pre - operative vitals were normal (heart rate: 100 beats/min, oxygen saturation: 100% and blood pressure: 96/65 mmHg). He was premedicated with IV fentanyl and anaesthesia was induced with IV propofol. Endotracheal intubation was facilitated with IV atracurium, while 100% oxygen and sevoflurane were used for maintenance. Surgery was uneventful and he was extubated after regaining full consciousness. Postoperative oxygen saturation was 100% and ABG was normal.

3. Discussion

Methaemoglobinaemia is one of the rare haemoglobinopathies that may present with 'dark - coloured arterial blood' with or without clinical signs and symptoms. Because of a mismatch between SaO₂ and SpO₂, which was not resolved after oxygen therapy, this anomaly alerted us to the possibility of abnormal hemoglobin The pathology implicated is oxidation of ferrous ion (Fe²⁺) of the Hb molecule to the ferric state (Fe³⁺) that hampers oxygen - binding capacity of Hb molecule and hence oxygen delivery to tissues. Normally, the amount of methaemoglobin is 70% result in circulatory collapse and acidosis and may be fatal.

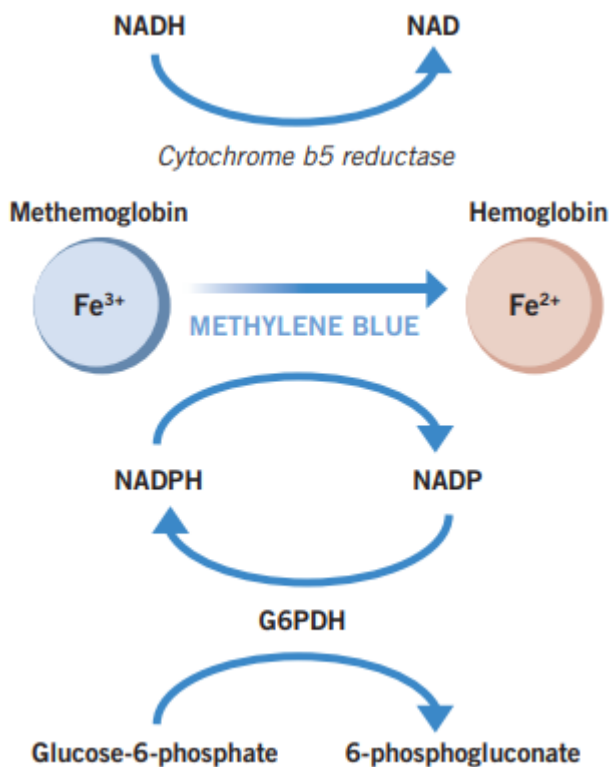


Figure 1: Pathways for methemoglobin reduction to hemoglobin. The main pathway is cytochrome b5 reductase. Drugs such as methylene blue also can reduce methemoglobin

4. Conclusion

In the present report, the child probably had congenital variety of the disease as evidenced by low levels of NADH methaemoglobin reductase enzyme and a family history of consanguinity with a normal elder sibling. Diagnosis is based on co - oximetric analysis of methaemoglobin percentage. Reduced levels of NADH methaemoglobin reductase enzyme are seen in the congenital variety. ABG is generally normal. The condition has to be distinguished from more common causes of cyanosis such as cardiorespiratory disease or sulfhaemoglobinemia (that is unresponsive to methylene blue).

References:

- [1] Mohorovic L, Materljan E, Brumini G. Consequences of methaemoglobinemia in pregnancy in newborns, children, and adults: Issues raised by new findings on methaemoglobin catabolism. *J MaternFetal Neonatal Med* 2010; 23: 956 - 9.
- [2] Baraka AS, Ayoub CM, Yazbeck - Karam V, Kaddoum RN, Gerges FJ, Hadi UM, et al. Prophylactic methylene blue in a patient with congenital methaemoglobinemia. *Can J Anaesth* 2005; 52: 258 - 61.
- [3] do Nascimento TS, Pereira RO, de Mello HL, Costa J. Methemoglobinemia: from diagnosis to treatment. *Rev Bras Anesthesiol*. 2008; 58 (6): 651 - 664.
- [4] Sharma VK, Haber AD. Acquired methemoglobinemia: a case report of benzocaine - induced methemoglobinemia and a review of the literature. *Clin Pulm Med*. 2002; 9 (1): 53 - 58.

- [5] US Environmental Protection Agency. Protect your home's water. www.epa.gov/privatewells/protect-your-homes-water#welltestanchor. Accessed October 25, 2019.
- [6] Nappe TM, Pacelli AM, Katz K. An atypical case of methemoglobinemia due to self - administered benzocaine. *Case Rep Emerg Med*. 2015; 2015: 670979.7. Barker SJ, Curry J, Redford D, Morgan S. Measurement of carboxyhemoglobin and methemoglobin by pulse oximetry: a human volunteer study. *Anesthesiology*. 2006; 105 (5): 892 - 897.
- [7] Boylston M, Beer D. Methemoglobinemia: a case study. *Crit Care Nurse*. 2002; 22 (4): 50 - 55.
- [8] Proxblue prescribing information. Shirley, NY: American Regent, Inc.; 2017. www.accessdata.fda.gov/drugsatfda_docs/label/2017/204630s0051bl.pdf. Accessed October 25, 2019.
- [9] Faust AC, Guy E, Baby N, Ortegon A. Local anesthetic - induced methemoglobinemia during pregnancy: a case report and evaluation of treatment options. *J Emerg Med*. 2018; 54 (5): 681 - 684