Unusual presentation of Acquired Methaemoglobinemia
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Citation

Abstract
We describe the case of a child with acquired methaemoglobinemia. As the history was strongly suggestive of foreign body aspiration, a rigid bronchoscopy was initially performed under general anaesthesia followed later by successful treatment with methylene blue.

The results of pulse oximetry in methaemoglobinemia are discussed and the reasons for the erroneous readings are explained.

INTRODUCTION
Pulse oximetry measurement of oxygen saturation in arterial blood is a widely used modern tool used for monitoring purposes in operation theatres, recovery rooms and intensive care units. The device uses plethysmography and light absorbance measurements at two different wavelengths to estimate oxygen saturation. The accuracy of this method may be affected by a number of factors; among them is the presence of abnormal haemoglobin, such as methaemoglobin and several therapeutic and diagnostic dyes.

We report the pulse oximetry analysis of a child who had high level of methaemoglobin (44.4%) leading to cyanosis. She underwent a rigid bronchoscopy, as she had a history which was suggestive of foreign body aspiration. Subsequently, a correct diagnosis of methaemoglobinemia was made followed by successful treatment with methylene blue.

CASE REPORT
A two years old female child weighing 10.5 kg was admitted with cyanosis. The parents described a history of a sudden bout of coughing while eating a lollipop followed by a brief episode of breathlessness, after which cyanosis was noticed. There was no history of any other illness except an upper respiratory tract infection for which she was receiving some drugs, the exact nature of which was not known to the parents. When examined, there was central as well as peripheral cyanosis. The respiratory rate was 34 breaths per min, pulse rate 112 beats per min. There was no evidence of stridor or respiratory distress and she had minimal fine crepitations. However air entry was bilaterally equal, sensorium was not altered and no other abnormality was found.

Her chest radiograph showed mild shift of the trachea to the left and bilateral hyperlucency. As there was definite history of aspiration, an arterial blood gas analysis was not done and the patient was taken up for rigid bronchoscopy on an emergency basis. In the operating room, a five lead ECG and the probe of an Ohmeda 3700 were attached. The saturation was seen to be 85 percent with the patient inhaling 100 percent oxygen. Bronchodilators and steroids were given. General anaesthesia was administered with nitrous oxide and halothane in oxygen. Relaxation was achieved with 2mg/kg suxamethonium and maintained with intermittent suxamethonium in 5 mg incremental doses. Ventilation was with Sander’s injector after the rigid bronchoscope was passed in. A thorough bronchoscopy revealed a few mucous secretions on the right side, which were sucked out. No foreign body was found and the procedure was terminated. Saturation remained 85 percent throughout the procedure. After spontaneous respiration resumed and consciousness returned, the saturation still remained 85 percent. The auscultatory findings were unchanged, as compare to preoperative. No fluctuation of any other vital parameters was seen during the procedure. The patient was still cyanosed and after observation in the operating room, the patient was transferred to the intensive care unit.

Radiograph of the chest was done which showed no change
from the radiograph done prior to the bronchoscopy. The arterial blood gas report done was as follows: PH = 7.39, PO$_2$ = 69 mmHg, PCO$_2$ = 27.1 mmHg, HCO$_3$ = 17.6 mmol/L, SaO$_2$ = 94.3%. As the cyanosis was deep and persistent despite the high level of oxygen saturation, a high index of suspicion for methaemoglobinemia was maintained and a blood sample was sent for methaemoglobin. Meanwhile, the child developed sudden high fever and altered sensorium. The blood pressure rose to 170/110 mmHg and sublingual Nifedepine 2.5 mg was given. The report which was subsequently received showed methaemoglobin of 44.4%.

Methylene blue was started in dose of 1 mg/kg given slowly intravenously and ascorbic acid 100 mg/kg/day were also given. Methaemoglobin level monitoring was continued on a daily basis. Subsequent reports of methaemoglobin showed a steady fall, till it was 6.7% on the fourth day (see Table 1).

Methylene blue was continued until the third day. Antibiotics and chest physiotherapy was given for treatment of the respiratory tract infection. By the second day, sensorium improved and blood pressure normalized. Cyanosis disappeared on the third day. The arterial blood gas report (on FIO$_2$ = 0.3) prior to discharge from intensive care unit, on the fourth day of admission of pH = 7.41, PO$_2$ = 157 mmHg, HCO$_3$ = 26.1 mmol/L, SaO$_2$ = 99.8%.

The patient was discharged from our hospital on the eighth day of admission.

**DISCUSSION**

Oxygen transport depends on the maintenance of intracellular haemoglobin in the reduced form (Fe$^{2+}$) form. When haemoglobin is oxidized to methaemoglobin, the heme becomes Fe$^{3+}$ and is incapable of binding oxygen. A small amount of haemoglobin autoxidises as the red blood cells circulate. Normally, methaemoglobin is reduced by cytochrome b$_5$.

Methaemoglobin forms 0.4% of the total Hb in normal circumstances. Congenital (due to cytochrome b$_5$ reductase deficiency) or acquired methaemoglobinemia (due to dapsone, phenacetin, sulfonamides, aniline dyes, lidocaine, nitrates and nitrates); results in clinically obvious cyanosis, if it exceeds 15g/L (1.5g/dl) i.e. 10 percent of the total haemoglobin with higher levels, patients become symptomatic and at 35 percent of methaemoglobin levels, the individual experiences headache, weakness and breathlessness. Levels in excess of 70 percent are incompatible with life. The structural configuration of methaemoglobin is very similar to that of oxyhaemoglobin.

Pulse oximetry by the standard Ohmeda Biox 3700 measures the relative absorption of red light at 660 nm and infrared light at 940 nm by HbO$_2$ and Hb and converts by computer the different amounts of light absorbed into SaO$_2$ and interprets the pulsatile component as pulse rate. The ratio of the absorption of 660 nm (maximum by reduced haemoglobin) to the absorption at 940 nm (maximum by oxyhaemoglobin) is converted to SaO$_2$ by an algorithm. According to this, the saturation approaches 100% as the ratio approximates a value of 0.4 (99% saturation at 0.43).

Methaemoglobin absorbs lights at 600 nm as well as 940 nm, so that at high levels of methaemoglobin, the ratio approaches unity. Algorithmically, this correlates to a value of 85 percent,$^2,10,11,15$ and this remains constant for a wide range of methaemoglobin. This at high levels of methaemoglobinemia, the pulse oximeter over estimates the saturation and at lower levels of methaemoglobinemia, it underestimates saturation. Methaemoglobin is measured in the laboratory by spectrophotometrically measuring the absorption of red light at 620-630 nm.$^{12}$

This child was being treated by a physician for her respiratory tract infection and the exact nature of drugs being given was unknown. It is possible that she was given sulfonamides. The cyanosis may have existed for a day or two, before it was actually noticed. Coincidentally this was after her bout of coughing while eating a lollipop and this resulted in a diagnosis of foreign body aspiration.

The unchanging (constant) reading of the pulse oximeter at 85% inspite of the wide variation in oxygenation that results during rigid bronchoscopy should have alerted us to the possibility of a different diagnosis, especially in view of the negative result of the bronchoscopy. The depth of the cyanosis did not correlate to the saturation of 85 percent on the pulse oximeter which was overestimating the saturation.
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The finding of a saturation of 94.3% on arterial blood gas estimation, by extrapolating the partial pressure of oxygen on the oxy-hemoglobin dissociation curve, raised a suspicion of methaemoglobinemia, which was confirmed by laboratory findings.

Methylene blue given intravenously caused a reversal of this condition, although the saturation by pulse oximetry remained at 85 percent for 48 hours more before steadily rising (although it was still less than saturation on the arterial blood gas report).

This condition can be more readily diagnosed by the presence of four wavelength oximetry, (which measures oxyhaemoglobin, reduced haemoglobin, methaemoglobin and carboxy-haemoglobin).

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