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# An Unusual Case of Methemoglobinemia

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## Article Info

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## Abstract

Methemoglobinemia is a condition in which an abnormal proportion of the iron in heme moiety of the hemoglobin is oxidized to the ferric state leading to impaired oxygen transport and anemic hypoxia and can cause tissue ischemia and death. A 21 year old male, known case of Hansen's disease was posted for surgicalneurolysis of ulnar nerve. His surgery was already postponed once as SpO2 waslow on the operation table. He then underwent complete systemic evaluation for the cause of low saturation but was found normal. On pulse oximetry it was seen that SpO2 was 91-93% on room air without any signs and symptoms of hypoxemia. This suggestedmethemoglobinemia due to treatment with dapsone which was being taken by the patient for treatment of Hansen's disease. The surgery was conducted uneventfully under general anesthesia. The diagnosis was confirmed by laboratory testing.

**Keywords:** Methemoglobinemia; Dapsone.

## Introduction

There are many causes of low oxygen saturation (SpO2) of hemoglobin in a patient apart from various cardiac and respiratory causes such as different type of hemoglobinopathies of which methemoglobinemia is the most common one. Methemoglobinemia is a condition in which an abnormal proportion of the iron in heme moiety of the hemoglobin is oxidized to the ferric state leading to impaired oxygen transport and anemic hypoxia and can cause tissue ischemia and death. It results most commonly from a toxic exposure, but in rare cases it can be hereditary [1]. About forty substances have been implicated in causing this condition, the most prominent being dapsone, nitrates, prilocaine, antimalarials and sulfonamids. Dapsone is used to treat several systemic inflammatory diseases, such as leprosy. 1 Treatment of leprosy with dapsone complicated by methemoglobinemia has an incidence of 5% [2,3].

## Case Report

A 21 year old male, known case of Hansen's disease was posted for surgical eurolysis of ulnar nerve in the department of orthopedics. He was on multidrug therapy which included rifampicin, clofazimine and dapsone for leprosy since one year. On preanesthetic evaluation he had good exercise tolerance, no other significant history but the treatment history was not given any importance by the resident anesthesiologist.

His surgery was already postponed once as SpO2 was low on the operation table. He then underwent complete systemic evaluation for the cause of low saturation but was found normal. He was therefore again posted for surgery. In the operation theatre, all routine monitors were attached. Onpulseoximetry it was seen that SpO2 was 91-93% on room air without any signs and symptoms of hypoxemia. An arterial blood gas (ABG) analysis done on room air was found to have oxygen saturation of 99.7%. On seeing the ABG report, consultant anesthesia took a detailed history of the patient and found out

that he was on dapsone treatment for leprosy which was missed by the residents. This suggested methemoglobinemia due to treatment with dapsone which was further established by laboratory investigations. The surgery was conducted uneventfully under general anesthesia. An ABG sample taken during surgery showed paO2 of 250 mmHg on FiO2 of 0.5.

A blood sample was sent for analysis and it was seen that patient had a methemoglobin level of 2g/dl (20%) level of methemoglobin (MetHb) confirming the diagnosis of methemoglobinemia.

## Discussion

There are two types of methemoglobinemia congenital and acquired: Congenital methemoglobinemia is characterized by diminished enzymatic reduction of methemoglobin back to functional hemoglobin [4,5]. Affected patients appear cyanotic but are generally asymptomatic. Most cases of the less common hereditary methemoglobinemias are due to homozygous or compound heterozygous deficiency (autosomal recessive) or in compound heterozygous cytochrome b5 reductase deficiency, which is primarily seen in sporadic cases. Another congenital cause of methemoglobinemia is hemoglobin M disease, which is due to mutations in a single globin gene (autosomal dominant) of either the alpha, beta, or rarely gamma globin gene. Deficiency of cytochrome b5 is the rarest form of congenital methemoglobinemia, and has been described in only one or two families. Acquired methemoglobinemia typically results from ingestion of specific drugs or agents that cause an increase in the production of methemoglobin.

Dapsone is a leprostatic agent commonly prescribed for the treatment of patients with leprosy, malaria, and a variety of blistering skin diseases, including dermatitis herpetic form is. Methemoglobinemia is a known adverse effect of dapsone use. Dapsone3 (4, 4′-diaminodiphenyl sulfone) is the parent compound of many sulfone medications, and has two primary toxicities: methemoglobinemia and hemolytic anemia.

Methemoglobinemia can be a fatal disease. Cyanosis is evident with only blood levels of 1.5g/dL of methemoglobin, in contrast to the 5g/dL of deoxygenated hemoglobin required to see hypoxia-related cyanosis. Levels between 20-45% are associated with dyspnea, lethargy, dizziness, lightheadedness, weakness and headaches. Levels> 45% are usually associated with impaired consciousness, and levels above 55% can cause seizures, coma and cardiac arrhythmias.

The diagnosis of methemoglobinemia is based on clinical symptoms and laboratory testing. Arterial blood gas analyses paired with oxygen saturation analysis by pulse oximetry are now considered the definitive measures for making a correct diagnosis of methemoglobinemia. Further evaluation of our patient's medical history revealed that he had been receiving treatment with dapsone for Hansen's disease. Our patient had no signs and symptoms of raised level of methemoglobinand it was diagnosed on the operation table after attaching the pulse oximeter.

For symptomatic patients, initial treatment includes administering oxygen. The treatment of choice is methylene blue [6,7]. It is administered typically in doses of 1 to 2 mg/ kg of body weight intravenously and is followed by rapid relief of symptoms [8,9]. Repeat doses may be indicated if symptoms persist, as may occur if there is continued absorption of the methemoglobin-inducer [8]. In cases of treatment failure with methylene blue, hyperbaric oxygen therapy and exchange transfusions can be considered.

## Conclusion

This report intends to show the importance of the awareness, diagnosisand treatment of this adverse effect of dapsone by the anesthesiologist.

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