Methemoglobinemia Diagnosis and Management

Methemoglobinemia is a disease caused by an excess of methemoglobin in the blood. Patients can present with cyanosis, respiratory distress, headache, and lethargy. Findings that may help make the diagnosis include "chocolate-colored blood", decreased oxygen saturation, and a normal partial pressure of arterial O₂ (PaO₂). This disease can be manage by identifying and avoiding triggers, methylene blue, vitamin C, and exchange transfusion.

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Clinical Features

Cyanosis

Cyan-Crayon

Cyanosis can be seen in patients with methemoglobinemia when methemoglobin makes up approximately 10% of total hemoglobin. Patients with oxygen saturation in the 80s (measured by pulse oximetry) are not typically cyanotic due to the oxygen saturation gap theory.

Respiratory Distress

Lungs Shooting Flare-gun

Respiratory distress can be seen in patients with methemoglobinemia when methemoglobin makes up approximately 30-50% of total hemoglobin.

Headache

Headache-egg-lump

Headache and other hypoxic symptoms can be seen in methemoglobinemia patients when methemoglobin makes up approximately 20% of total hemoglobin.

Lethargy

Leather-jacket

Lethargy or loss of consciousness can be seen in methemoglobinemia patients when methemoglobin makes up approximately 30-50% of total hemoglobin. If it is near 50%, severe symptoms may occur, such as arrhythmias, seizures, metabolic acidosis, and coma. Mortality increases as binding approaches 70%.

Diagnosis

"Chocolate-colored Blood"

Chocolate-bar Blood

Normally, the color of blood is red due to the presence of oxygen bound to hemoglobin. Patients with methemoglobinemia have a reduced oxygen-carrying capacity in their blood. This will cause the patient's blood to appear dark red to brown. This can be easily seen if the blood is dripped on a piece of white gauze.

Decreased Oxygen Saturation

Down-arrow Percent O2-tank

Pulse oximetry in patients with methemoglobinemia will erroneously reveal a number around 85%. This occurs due to the high absorption of light by methemoglobin. Under normal conditions, pulse oximetry measures the ratio of oxygenated and deoxygenated hemoglobin by measuring the absorbance of light. Methemoglobin, however, absorbs light differently than both oxygenated and deoxygenated hemoglobin. Thus, the pulse oximeter will reveal a falsely elevated oxygen saturation despite the actual oxygen saturation. For example, a methemoglobin level of 5% and 40% will both present on pulse oximetry as an O2 saturation of ~85%. Oxygen saturation on blood gas analysis can be falsely high as it is based on the PaO2, which reflects plasma oxygen content. Because of this, patients can present with a saturation gap. This is also referred to as "refractory hypoxemia."

Normal Partial Pressure of Arterial O₂ (PaO₂)

Normal-sign Partial Pressure-Gauge Artery-Archer with O2-Tank

Normal PaO2 is commonly seen in patients' blood gas analysis despite cyanosis. This finding suggests methemoglobinemia.

Management

Identify Triggers

Magnifying-glass Identifies Triggers

The first step in treating methemoglobinemia is to identify triggers and stop the inciting agent. Potential triggers include dietary nitrates and drugs such as nitroglycerin, dapsone, and lidocaine.

Methylene Blue

Metal-flail Blue

Methylene Blue is a drug that reduces methemoglobin via NADPH. It will convert methemoglobin to hemoglobin. It is used as the primary emergency treatment for symptomatic methemoglobinemia and should be used with caution in patients with G6PD deficiency since it depletes NADPH reserves and may cause oxidative crisis in G6PD deficiency patients.

Vitamin C

Viking Orange

Vitamin C can be used for the treatment of methemoglobinemia. It is indicated if methylene blue is not available, ineffective, or not recommended.

Exchange Transfusion

Exchanging Transfusion-IV

An exchange transfusion can also be initiated If methylene blue does not work or is not recommended in a patient. It works by replacing abnormal methemoglobin with normal hemoglobin.
