Ferric form of heme to the ferrous form.

NADH mutation, inherited in an autosomal recessive pattern, results in a deficiency of Hereditary Methemoglobenia is rare and few cases have been reported. A genetic

Symptoms

metHb in the blood exceeds 2g/dL as well as impairing the release of O₂ hemoglobin dissociation curve therefore decreasing the oxygen carrying capacity as a result of a left shift of the Methemoglobin

DISCUSSION:

We decided to proceed, assuming a diagnosis of hereditary methemoglobinemia. A standard mask induction was uneventful. In addition to an intravenous line, an arterial line was placed for frequent blood gas measurements. Twenty minutes after induction, an arterial blood gas (ABG) measurement showed an OxyHgb of 60% and a MetHgb of 25.9% on 100% FiO₂.

Subsequent blood gas and SaO₂ measurements was similar (table 1). The remainder of the anesthetic was uneventful.

Methemoglobin

< 1.5 g/dL 10% None

1.5-3.0 g/dL 10-20% Cyanotic skin discoloration

3.0-4.5 g/dL 20-30% Anorexia, lightheadedness, headache, tachycardia

4.5-7.5 g/dL 30-50% Fatigue, confusion, dizziness, tachypnea, tachycardia

7.5-10.5 g/dL 50-70% Coma, seizures, arrhythmias, acidosis

> 10.5 g/dL >70% Death

* Assumes hemoglobin = 15 g/dL. Patients with lower hemoglobin concentrations may experience more severe symptoms for a given percentage of methemoglobin level.

* Patients with underlying cardiac, pulmonary, or hemolytic disease may experience more severe symptoms for a given methemoglobin concentration.

Summary of Patient's Perioperative Data

<table>
<thead>
<tr>
<th>Methemoglobin</th>
<th>% Total Hemoglobin*</th>
<th>Symptoms*</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 1.5 g/dL</td>
<td>10%</td>
<td>None</td>
</tr>
<tr>
<td>1.5-3.0 g/dL</td>
<td>10-20%</td>
<td>Cyanotic skin discoloration</td>
</tr>
<tr>
<td>3.0-4.5 g/dL</td>
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</tr>
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<td>&gt; 10.5 g/dL</td>
<td>&gt;70%</td>
<td>Death</td>
</tr>
</tbody>
</table>

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It is associated with or without neurologic symptoms, type I and II, respectively. Abnormal hemoglobin variants referred to hemoglobin M, inherited in an autosomal recessive pattern results in inefficient reduction of iron to the ferrous form. Cyanosis in consecutive generations, as in the case of our patient, suggests the presence of hemoglobin M.

Perioperative treatment is often not warranted in asymptomatic hereditary methemoglobinemia. Both Methylene blue and Ascorbic Acid can be used in the treatment of symptomatic patients in most cases.

Exchange transfusion and/or hyperbaric oxygen are used in critical situations[12]

CONCLUSION:

Hereditary Methemoglobinemia is an important cause of cyanosis, and early detection and diagnosis is critical. Perioperative management of these patients requires O₂ supplementation, frequent co-oximetry monitoring, avoiding oxidizing drugs and treatment with methylene Blue or Exchange transfusion. Prior knowledge of this condition can assist with management and avoid delay in treatment in the perioperative setting.

REFERENCES

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3. Crit Care Med. 2003; Apr;31(4):1110-4