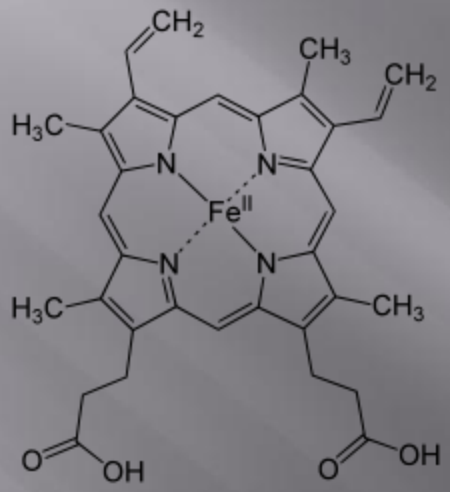
METHEMOGLOBINEMIA



Peter Sambatakos
PGY1
4/20/12

1) 20 y/o mother with a h/o canker sores applied her Orabase gel (20% benzocaine) to her 12 month old previously healthy infant for concern of teething pain. Soon thereafter, infant became "lethargic" and had a bluish hue according to mother. She brought him in to the ER for evaluation. On exam, infant has a HR of 156, T 37.5, BP 105/78, RR 52, O2 86% . What would be your next step in management?

A) ABG

B) CBC, BMP

C) oxygen therapy

D) methylene blue

E) hyperbaric oxygen

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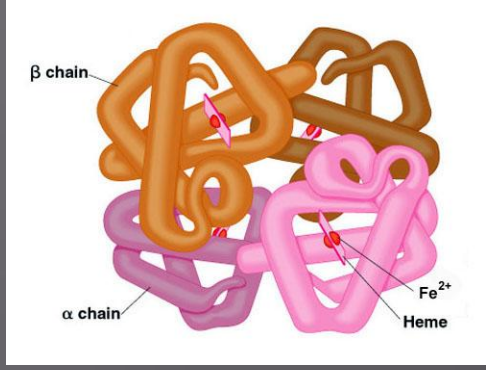

- Methemoglobin has an oxidized ferric iron (Fe +3) rather than the reduced ferrous form (Fe 2+) found in hemoglobin. This structural change is responsible for methemoglobin's inability to bind oxygen. In addition, ferric iron has slightly greater affinity for oxygen due to its chemical structure, thus shifting the oxygen dissociation curve of partially oxidized hemoglobin molecules to the left, resulting in decreased release of oxygen in tissues. The findings of anemia and cyanosis despite oxygen treatment result from both of these effects

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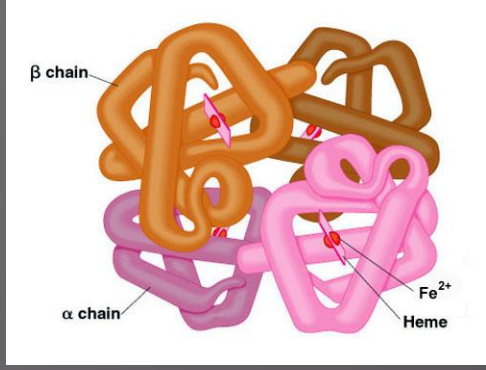

- Symptoms are proportional to the level of methemoglobin.
  - Less than 10% methemoglobin – No symptoms
  - 10-20% methemoglobin – Skin discoloration only (most notably on mucus membranes)
  - 20-30% methemoglobin – Anxiety, headache, dyspnea on exertion

continued

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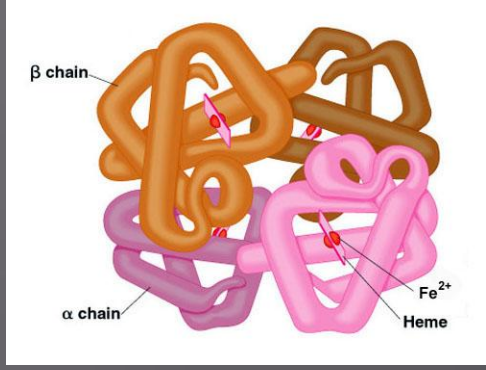

- 30-50% methemoglobin – Fatigue, confusion, dizziness, tachypnea, palpitations
- 50-70% methemoglobin – Coma, seizures, arrhythmias, acidosis
- Greater than 70% methemoglobin – Death

- The presence of methylene blue allows NADH reductase to function at an increase rate.

2) 100% O2 therapy is initiated and patient's O2 remains at 86%. ABG reveals a PaO2 of 98 mmHg. Methemoglobinemia is suspected and methylene blue 1 mg/kg is administered IV. Over the next several hours the patient now looks like:

- UA shows large RBCs. CBC reveals a Hgb of 10.8 (initial Hgb was 12.3) with Heinz bodies on peripheral smear. What is the etiology of patient's current symptoms?

A) Hypersensitivity reaction to methylene blue

B) G6PD deficiency

C) Sickle cell disease

D) Autoimmune hemolysis

E) Thalassemia

- UA shows large RBCs. CBC reveals a Hgb of 10.8 (initial Hgb was 12.3) with Heinz bodies on peripheral smear. What is the etiology of patient's current symptoms?

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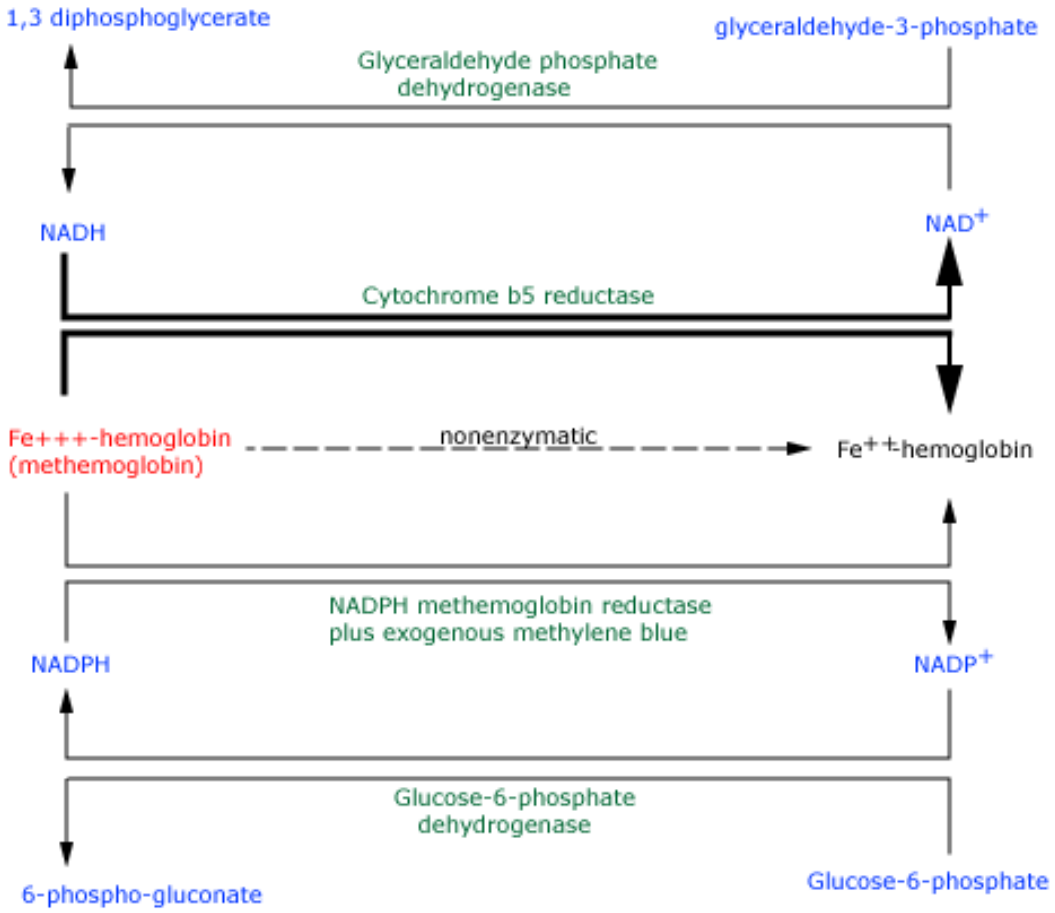
Metabolic pathways for reduction of methemoglobin



The major pathway for methemoglobin reduction is via cytochrome b5 reductase (thick arrows). An alternative pathway, which requires an exogenous electron acceptor such as methylene blue, is via NADPH methemoglobin reductase. Only a small amount of methemoglobin is reduced via nonenzymatic pathways (dashed arrow).

References

- http://cdn.womentribe.com/wp-content/uploads/2011/11/Methemoglobinemia-Syndrome-in-babies.jpg&imgrefurl=
- http://emedicine.medscape.com/article/204178-overview
- http://www.amazon.com/Orabase-Paste-20-Benzocaine/dp/B0000Y3I46
- http://www.medscape.com/viewarticle/410479_2
- http://www.uptodate.com/contents/clinical-manifestations-of-glucose-6-phosphate-dehydrogenase-deficiency ?source=preview&anchor =H5& selectedTitle=2~150#H1
- http://www.uptodate.com/contents/autoimmune-hemolytic-anemia-in-children?source=preview&anchor=H10&selectedTitle=4~150#H10
- http://www.womentribe.com/pregnancy-parenting/methemoglobinemia-syndrome-in-babiescausessymptomstreatment.html&usg
- *Methylene Blue-induced Hyperbilirubinemia in Neonatal Glucose-6-phosphate Dehydrogenase (G6PD) Deficiency.* Theresa W. Gauthier. The Journal of Maternal-Fetal Medicine 9:252Ð254 (2000)
- *Recognition and Management of Methemoglobinemia and Hemolysis in a G6PD-Deficient Patient on Experimental Anticancer Drug Triapine.* Lynda M. Foltz,1 Bakul I. Dalal,2* Louis D. Wadsworth,3 Raewyn Broady,1 Kim Chi. American Journal of Hematology 81:210–211 (2006)