

## Anesthesia for Esophagogastroduodenoscopy in a patient with Santa Ana Hemoglobinopathy

Medhat Hannallah<sup>1</sup>, Anna Budde<sup>2</sup>

<sup>1</sup>Department of Anesthesiology, Georgetown University Hospital

<sup>2</sup>Georgetown University School of Medicine, Washington, DC, USA

### \*Corresponding Author:

Name: Anna Budde

Email: [amb328@georgetown.edu](mailto:amb328@georgetown.edu)

**Abstract:** Santa Ana hemoglobinopathy is a rare hemoglobinopathy which has been reported in the literature. We report a patient with Santa Ana hemoglobinopathy who presented for esophagogastroduodenoscopy (EGD). Santa Ana hemoglobinopathy is a genetic defect in the hemoglobin molecule which makes it unusually susceptible to thermal degradation. As the hemoglobin molecule denatures it changes conformation which means that pulse oximeters are not able to sense the hemoglobin and detect whether or not it is carrying oxygen. Such patients therefore have aberrantly low pulse oximeter measurements. If patients with this rare disorder are to be anesthetized, the pulse oximetry readings will not be useful to determine true oxygen saturation and alternative monitoring such as arterial line placement and repetitive arterial blood gas measurement will be necessary.

**Keywords:** Santa Ana hemoglobinopathy, esophagogastroduodenoscopy, gastroesophageal reflux.

### INTRODUCTION

Santa Ana hemoglobinopathy is a rare condition associated with aberrantly low pulse oximetry readings. When anesthetizing patients with such hemoglobinopathy, monitoring of ventilation has to be conducted without the benefit of continuous pulse oximetry.

We report a patient with Santa Ana hemoglobinopathy who presented for esophagogastroduodenoscopy (EGD). This procedure is commonly performed during propofol anesthesia without endotracheal intubation. It is also a procedure that has the potential for causing upper airway irritation, laryngospasm, and reduction in arterial oxygen saturation.

### CASE REPORT

A 45 yr-old male with Santa Ana hemoglobinopathy presented for EGD for evaluation of symptoms of gastroesophageal reflux. The only manifestation of his condition was pulse oximetry reading of  $\pm 85\%$  with or without supplemental oxygen. He was 68 cm tall, weighed 72 kg, and had a Mallampati class 2 airway. After consideration of different options and in view of the short duration of the procedure and the normal airway, a routine anesthetic technique was used. Monitoring included non-invasive blood pressure measurement, electrocardiography, capnography, precordial stethoscope, and pulse oximetry. The patient was placed in the left lateral decubitus position. Supplemental O<sub>2</sub> was given via nasal cannula. A total propofol dose of 2.0 mg/kg was

slowly administered until there was no response to oropharyngeal stimulation. Anesthesia was maintained with propofol 200 mcg/kg/min. The pulse oximeter reading remained in the mid-80's throughout the uneventful 5 minute procedure.

### DISCUSSION

Hemoglobinopathies are defects in the hemoglobin molecule that hinder its ability to carry oxygen effectively. Approximately 7% of the world population is affected by one of these genetic disorders making them a relatively common problem [2]. Hemoglobinopathies can be classified into several different types including thalassemias, disorders with abnormal hemoglobin, and unstable hemoglobinopathies. Unstable hemoglobinopathies are congenital diseases which present with hemolytic anemia, splenomegaly, and reticulocytosis. Often hemoglobin precipitates and is degraded within the red blood cell leading to a decrease in oxygen carrying capacity [3].

Santa Ana hemoglobinopathy represents a rare variant of unstable hemoglobinopathy which presents with Heinz body anemia, pigmenturia, and hemolytic anemia early in life. The hemoglobin in this disorder is unusually sensitive to thermal denaturation which is thought to be the cause of the hemoglobin's instability. The treatment for this disorder is splenectomy which usually occurs early in life. Laboratory studies performed in a family with Santa Ana hemoglobinopathy show that these patients are no longer anemic after splenectomy (with an average

hematocrit of 39% among the three family members studied) but all three patients' erythrocytes had abnormal osmotic fragility and were prone to thermal denaturation [1].

Patients with hemoglobinopathies often have abnormal readings on pulse oximetry. Pulse oximeters rely on the light absorbance of oxygenated and deoxygenated hemoglobin to estimate the arterial blood saturation. The wavelengths in a pulse oximeter are set to 660 nm for deoxygenated hemoglobin and 940nm for oxygenated hemoglobin. Variant hemoglobins present in patients with hemoglobinopathies may prevent the accurate measurement of arterial blood saturation because these variant hemoglobins may absorb light at slightly different wavelengths than are measured by the standard pulse oximeter [4].

The result of the variant hemoglobin absorbing light at slightly different wavelengths may be an underestimation of the arterial blood saturation by the pulse oximeter which explains the aberrantly low pulse oximeter readings that can occur in patients with hemoglobinopathies. It is also possible for variant hemoglobins to have lower oxygen affinity than normal meaning that the low oxygen saturation measured by the pulse oximeter is an accurate reflection of low arterial blood saturation. Therefore, a patient with Santa Ana hemoglobinopathy may have aberrantly low pulse oximetry measurements because the Santa Ana variant hemoglobin differs structurally from normal hemoglobin and absorbs light at a different wavelength which is not measured by a pulse oximeter. It is also possible that these patients have accurately low pulse oximetry measurements because Santa Ana hemoglobin is unstable and does not have a normal oxygen carrying capacity and the patients truly have decreased arterial oxygen saturation.

## CONCLUSION

Pulse oximetry is an essential monitor in modern anesthesia practice. In its absence other monitoring techniques can provide indirect assessment of oxygenation. Capnography can provide very useful monitoring of ventilation. However, it is not consistently reliable in non-intubated patients who may be mouth breathers. Mouth breathing is common during EGD as the gastroscope functions as an oral airway which patients may preferentially breathe around. In this situation reliance must be upon clinical assessment of ventilation via auscultation, inspection of the chest excursions, and feeling for air exchange in front of the patient's mouth. If airway difficulty is anticipated elective endotracheal intubation should be considered. For major surgery arterial line placement and frequent blood gas analysis should be helpful.

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