AEJ ANESTHESIA EJOURNAL
Volume 9- No. 7 2021

Decreased Pulse Oximetry Readings in Asymptomatic Patient with Hemoglobin Grifton

Anne Marie Serrano[1], Christian R Serrano[2], and Monica Jenschke[3]

Affiliation:

[1] *Anne Marie Serrano, BSN, RN, Texas Christian University, School of Nurse Anesthesia, Fort Worth, TX*
[2] *Christian R Serrano, MS, BS, Department of Biology, University of Texas at San Antonio, San Antonio, TX*
[3] *Monica Jenschke, PhD, CRNA, Texas Christian University, School of Nurse Anesthesia, Fort Worth, TX*

Grant/Financial Support:
None

Biographical data:
Elaine Juch is a senior RRNA at Texas Christian University in Fort Worth. She would like to acknowledge the work of Courtney McNeill, CRNA, preceptor for guidance during this unique case.

KEYWORDS: Hemoglobin Grifton, hemoglobinopathies, pulse oximetry

Abstract

Hemoglobin Grifton presents as a decreased oxygen saturation as measured by pulse oximetry (SpO_2) reading despite a normal arterial oxygen saturation (SaO_2) in an asymptomatic patient. Hemoglobin Grifton is caused by a mutation on an alpha chain of hemoglobin at codon 87, which affects the absorption wavelength of its oxyhemoglobin. A standard pulse oximeter is designed to measure the absorption of light at specific wavelengths and perceives the hemoglobin Grifton oxyhemoglobin as a deoxyhemoglobin. In a patient with a known variant hemoglobin, ordering additional diagnostic or therapeutic testing can increase stress for a patient requiring routine medical interventions or emergencies. Careful evaluation of each individual case is warranted prior to proceeding with an anesthetic. Previous case studies reported variant hemoglobinopathies, specifically on the alpha chain, that produce a false low SpO_2 reading. In these case studies, the arterial blood gas resulted in normal oxygen saturation. Another case study reported that the issue with hemoglobin Grifton was the oxyhemoglobin Grifton absorbed light at approximately 740 nm. The light absorption on normal oxyhemoglobin is at a wavelength of 940 nm. Because of this, the hemoglobin Grifton is detected by the pulse oximeter monitor as deoxyhemoglobin. Standard transcutaneous pulse oximeters do not consider the different absorbance spectra of variant hemoglobinopathies where the oxyhemoglobins may be absorbed at lower spectrums. In all the literature reviewed, patients went through a myriad of unnecessary diagnostic tests to figure out the cause of the hypoxia. Some patients were even prescribed treatment based on this false hypoxia. This results in unnecessary additional expenses for treatment and can also cause stress for patients. The patient in this case study presented with a baseline SpO_2 of 84% with a known diagnosis of hemoglobin Grifton. The surgeon was unable to do this procedure in the clinic due to the child's agitated behavior and general anesthesia was required. The decision was made to forgo invasive testing due to her known condition and the unnecessary stress it would add to the patient.

Decreased Pulse Oximetry Readings in Asymptomatic Patient with Hemoglobin Grifton

Anne Marie Serrano, BSN, RN, Texas Christian University

Introduction

- Hemoglobin Grifton is caused by a mutation on the *HBA1* gene at codon 87 altering an alpha chain of the hemoglobin molecule (histidine → proline) (Figure 1).[1,2]
  - This specific location seems to affect the absorption wavelength of the oxyhemoglobin causing a false low oxygen saturation by pulse oximetry (SpO_2), yet the person has a normal arterial oxygen saturation (SaO_2).[1]

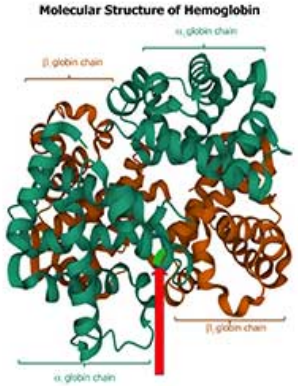

Figure 1. Molecular structure of oxyhemoglobin. The image is rotated to expose location of codon 87, highlighted with arrow.[3,4]

- Previous studies report patients with this similar type of hemoglobinopathy have a false low SpO_2, but have a normal SaO_2, PaO_2, and P_{50} when breathing room air.[1]
- A patient with a known hemoglobin variant that causes a falsely low SpO_2 with normal SaO_2 and is asymptomatic may not need invasive monitoring or extra testing for routine medical interventions.[1,2,5,6]

Case Report

- A 5-year-old female requiring general anesthesia for foreign object removal in ear.
- The surgeon was unable to do the procedure in the office due to the patient being uncooperative and easily agitated when examined.
- Past medical history: hemoglobin Grifton, hip anteversion
- No previous anesthetic or lab work
- Pre-anesthetic evaluation: baseline SpO_2 84% on room air
- The patient was visibly stressed and became increasingly agitated when attempting pre-anesthetic assessment.
- Steps taken to reduce agitation: having the parents involved in introducing a "play" mask; limiting the number of people to avoid overstimulation; and 10 mg midazolam, oral, pre-operatively
- Clinical observations: pink color, warm to touch, normal capillary refill time, and no signs of respiratory distress.
- The decision to forgo invasive monitoring was based on the patient's stressed behavior, a normal physical assessment, and estimated time of the procedure to be less than 15 minutes.

Anesthesia Management

Intra-operative

- Patient induced via mask with O_2/N_2O and sevoflurane
- Patient maintained spontaneous respirations
- SpO_2 readings remained approximately 84% despite administration of supplemental oxygen

Post-operative

- Patient transferred to PACU with blow-by oxygen
- No anesthetic complications during procedure
- Initial SpO_2 reading in PACU was 86%
- Patient was observed to be pink in color, warm to touch, and in no respiratory distress
- Handoff given to PACU RN about known history of hemoglobin Grifton and baseline SpO_2 of 84% on room air

Background

- Common causes of low or spurious pulse oximeter readings include hypoxia, ambient light interference, cold digits, poor perfusion states, excessive movement, IV pigmented dyes, fingernail polish, and inherited forms of abnormal hemoglobin.[7]

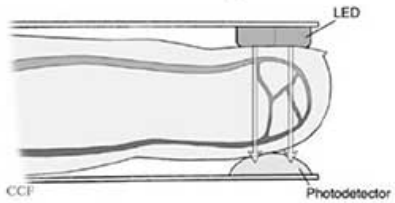

Figure 2. A standard transcutaneous pulse oximeter emits light across a vascular bed and calculates the proportions of oxyhemoglobin and deoxyhemoglobin based on their different absorption spectra.[1,2,5,6,8]

- Hemoglobin variants can interfere with the ability of the transcutaneous pulse oximeter to detect the light absorption of oxyhemoglobin and deoxyhemoglobin and may result in a false low SpO_2 reading (Table 1).[1,2,5,6,9]

Hemoglobinopathy	SpO_2	SaO_2
Hb Grifton*†	low	normal
Hb Bonn*†	low	normal
Hb Cherverly*	low	normal
Hb Koln*	low	normal
Hb Lansing*†	low	normal
Hb Titusville	low	normal
Hb Delaware	low	normal
Hb Hammersmith	low	normal
Hb Okazaki	low	normal
Hb Regina	low	normal
Hb Bassett	low	low
Hb Rothschild	low	low
Hb Canbiere	low	low
Carboxyhemoglobin	high	low
Methemoglobin	high	low
* Affects absorption spectrum		
† Mutation at codon 87		

- Hemoglobin Grifton oxyhemoglobin has increased light absorption at wavelengths from 600 to 740 nm. Deoxyhemoglobin absorbs light at 660 nm. Hemoglobin Grifton is perceived as deoxyhemoglobin by a standard pulse oximeter (Figure 3).[1]

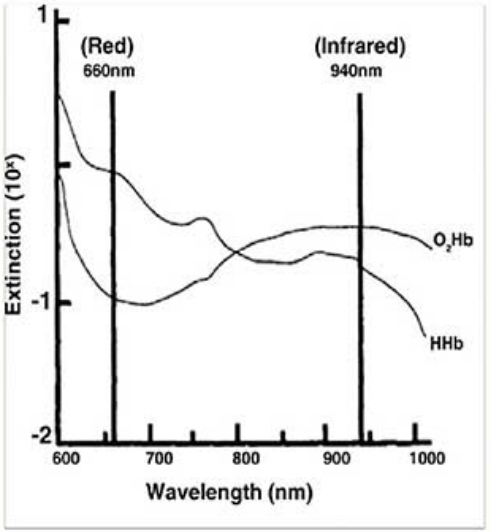

Figure 3. Hemoglobin extinction curve of normal adult oxyhemoglobin and normal adult deoxyhemoglobin.[6]

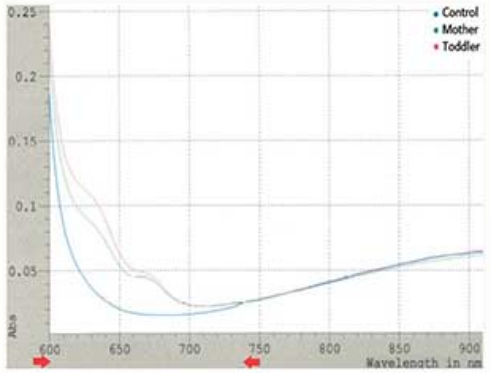

Figure 4. Spectrophotometry. Hb Grifton has increased absorption of light at wavelengths 600 to 740 nm compared to normal oxyHb controls, denoted between the red arrows.[1]

- In a patient with a known hemoglobinopathy where there is a discrepancy between SpO_2 and SaO_2, invasive testing may be unnecessary and cause more stress and anxiety for the patient.[1,2,5,6]

Conclusion

- A patient with a variant hemoglobin, such as hemoglobin Grifton, may present with low SpO_2 yet have a normal SaO_2.
- The patient does not have a true hypoxemia.
- The false hypoxemia is due to the transcutaneous pulse oximeter recognizing hemoglobin Grifton's oxyhemoglobin as deoxyhemoglobin based on the monitor's design.[7]
- Invasive testing or monitoring may be unnecessary and can cause more stress and anxiety for the patient.[1,2,5,6]

Recommendations and Suggestions

- Other variant hemoglobins with mutations at codon 87 of the alpha-globin chain suggest that this region has an impact on the oxyhemoglobin absorption spectrum and should be investigated further.[1,2,5]
- Use clinical observation to check if the patient is being adequately oxygenated. These observations include assessment of skin color and temperature; nail-bed perfusion signs; depth and rate of respirations; and upper airway patency.[10,11]
- In a patient who presents with a low pulse oximeter measurement with no known history of a variant hemoglobinopathy, consider using the algorithm for evaluation of low SpO_2 (figure 5).
- In a patient with a known hemoglobin variant that causes a low SpO_2 and normal SaO_2, additional invasive monitoring or testing may be unnecessary for routine medical interventions.[2,5-7]

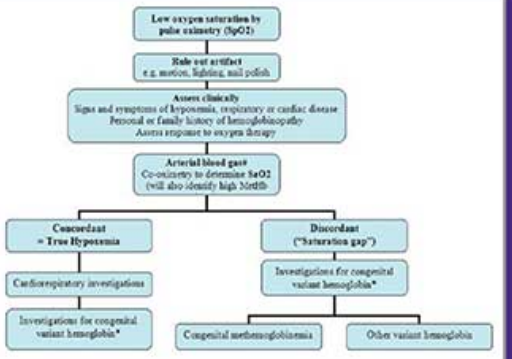

Figure 5. Algorithm for evaluation of low SpO_2. Arterial blood gas should be done on room air and with simultaneous SpO_2 measurement.[2]

References

1. Waters AM, Caboot JB, Verhovsek MM, Harper DP, Forouhar MA. Hb Grifton [α87(F8)His→Pro; HBA1: C.261A > C (or HBA2)] causes abnormal pulse oximetry measurements. *Hemoglobin*. 2016;40(4):257-259. doi:10.1080/03630269.2016.1174872
2. Verhovsek M, Henderson MP, Cox G, Luo HY, Steinberg MH, Chui DH. Unexpectedly low pulse oximetry measurements associated with variant hemoglobins: a systematic review. *Am J Hematol*. 2010;85(11):882-885. doi:10.1002/ajh.21810
3. Gumpper RH, Terrell JR, Luo M. 6BB5 Human oxy-hemoglobin. Research Collaboratory for Structural Protein Data Bank (RCSB PDB). 3D view image. January 10, 2018. Accessed April 4, 2021. doi:Bioinformatics10.2210/pdb6BB5/pdb; https://www.rcsb.org/structure/6BB5
4. Tomă JR, Gumpper RH, Luo M. Hemoglobin crystals immersed in liquid oxygen reveal diffusion channels. Biochem Biophys Res Commun. 2018;495(2):1858-1863. doi:10.1016/j.bbrc.2017.12.038
5. Zur B, Homung A, Breuer J, et al. A novel hemoglobin, Bonn, causes falsely decreased oxygen saturation measurements in pulse oximetry. *Clin Chem*. 2008;54(3):594-596. doi:10.1373/clinchem.2007.095158
6. Guler S, Brunner-Agten S, Bartenstein S, et al. Oxygen saturation of 75%, but no symptoms. *Respiration*. 2016;92(6):420-424. doi:10.1159/000451030
7. Sinex JE. Pulse oximetry: principles and limitations. *Am J Emerg Med*. 1999;17(1):59-67. doi:10.1016/s0735-6757(99)90019-0
8. Mace SE. The fifth vital sign: pulse oximetry in noninvasive respiratory monitoring. *Pediatr Emerg Med Rep*. March 1, 2005. https://www.reliasmedia.com/newsletters/25/issues/41978
9. Chan ED, Chan MM, Chan MM. Pulse oximetry: understanding its basic principles facilitates appreciation of its limitations. *Respir Med*. 2013;107(6):789-799. doi:10.1016/j.rmed.2013.02.004
10. Bozimowski G. Clinical monitoring II: respiratory and metabolic systems. In: Nagelhout JJ, Plaus KL, eds. *Nurse Anesthesia*. 5th ed. Elsevier; 2014:313-324.
11. Walsh BK, Smallwood CD. Pediatric oxygen therapy: a review and update. *Respir Care*. 2017;62(6):645-661. doi:10.4187/respcare.

- *Note:* The TCU IRB determined the case report scholarly endeavor does not require IRB approval and oversight.