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Pulmonary Medicine

SPURIOUS LOW OXYGEN SATURATION ON PULSE OXIMETRY IN AN ADULT PATIENT WITH HAEMOGLOBINOPATHY

Dr. Abraham Koshy* — Department Of Pulmonary Medicine, B. J. Medical College, Ahmedabad
*Corresponding Author

Dr. R. N. Solanki — Department Of Pulmonary Medicine, B. J. Medical College, Ahmedabad

Dr. Pyarpinkesh N. Rathva — Department Of Pulmonary Medicine, B. J. Medical College, Ahmedabad

ABSTRACT The use of pulse oximetry to estimate oxygen saturation in a continuous, accurate and non-invasive fashion has transformed vital monitoring in medical field . However, there are situations where there could be significant variance between oxygen saturation measurements with pulse oximeter (SpO2) and actual arterial oxygen levels (PaO2). In such scenarios, the possibility of a variant haemoglobin as the etiology exists. The finding is usually incidental, and patients themselves are often asymptomatic. There exists inadequate knowledge of principles and implications of pulse oximetry among health care providers. Awareness of these variants can help spare physicians and patients stress and unnecessary medical investigations. Further research into magnitude and types of variant haemoglobins in Indian population can help in proper diagnosis and management of such patients. Here, we present a case report of a patient presenting with low oxygen saturation who underwent extensive workup and was found to have a type of variant haemoglobin.

KEYWORDS : Spurious pulse oximetry, Haemoglobinopathy, Variant Haemoglobins

Introduction

The use of Pulse oximetry to estimate oxygen saturation in a continuous, accurate and non-invasive fashion has transformed medical monitoring, and is even referred to as the fifth vital sign [1]. Pulse oximetry can alert if something's amiss with the cardio-pulmonary systems and prompt the need for further investigation. In fact, measurement of oxygen saturation via pulse oximeter (SpO2) has de facto become the universal measure of arterial oxygen levels (PaO2).

In some situations however, the use of a pulse oximeter to measure oxygen saturation may not be accurate (Table 1). While in other settings, patients may unexpectedly be found to have low oxygen saturation but have normal arterial oxygen saturation levels on arterial blood gas analysis.

Pulse oximetry is based on the principle that Oxy-Haemoglobin (O2Hb) absorbs more near-Infra Red light than Deoxy Haemoglobin (HHb) and that HHb absorbs more red light than O2Hb [2,3,4]. It depends on a normal oxygen-haemoglobin dissociation curve. In presence of a variant haemoglobin however, there can be a discordance between SpO2 and PaO2 measurements [5, 6]. In the majority, the finding is usually incidental, and the patients are often healthy and asymptomatic. Therefore, knowledge of these variant haemoglobinopathies is helpful in alleviating stress of physicians in finding a proximate cause while sparing patients unnecessary medical investigations [7]. Here, we present a case report of a patient presenting with low oxygen saturation who underwent extensive workup and was found to have alpha variant haemoglobin.

Case Report

A 32 year old female was referred to our department for persistent complaints of breathlessness on exertion and headache since one year after child birth. There was no history of fever, cough, sputum production, loss of weight or appetite. Patient denied palpitations, syncope, associated chest pain, and pedal edema. There was no history of tobacco use or exposure to noxious stimuli.

On physical examination, pulse and blood pressure were normal, but oxygen saturation was 85% at rest on room air as obtained via pulse oximeter. Patient was admitted and evaluated. She had only a minor improvement of SpO2 readings with 6L of supplemental oxygen via facemask. Chest x-ray was obtained and was normal. Lung fields were clear. Heart sounds were regular, without murmur, gallops, or heaves. Abdominal examination was normal. An arterial blood gas (ABG) analysis showed a partial pressure of oxygen (paO2) of 123 mmHg, partial pressure of carbon dioxide (paCO2) 33mm Hg and pH 7.51 and an oxygen saturation of 98%. Due to persistent low SpO2 readings, further workup was initiated.

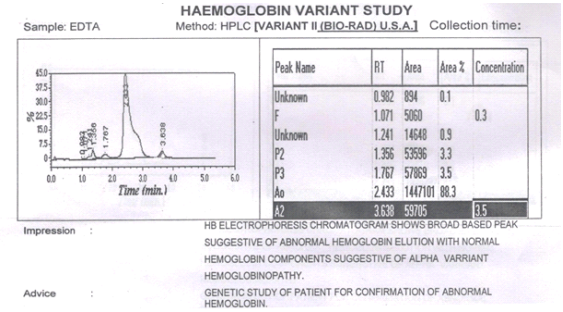
Blood reports were normal except a low haemoglobin level of 9.38mg/dl. ECG, 2D echo and contrast 2D echo were found to be normal. Pulmonary function tests showed no obvious derangement. High Resolution CT chest was normal, so was a CT pulmonary angiogram. Doppler studies and D dimer levels were normal. Cardiology and Haematology consultations were done. A haemoglobin electrophoresis was advised which showed Alpha variant haemoglobinopathy (Figure 1.)

Patient had a two-week long hospitalization for workup of hypoxia during which she had occasional complaints of shortness of breath, more on exertion while vitals and respiratory and cardiovascular examination remained normal.

Familial screening study showed alpha haemoglobinopathy in the patient's mother as well as in her male child. Interestingly, her husband's study showed findings suggestive of beta thalassemia trait.

Patient was discharged and advised follow up as an outpatient with hematology department. The patient was advised further genetic study to ascertain type of variant haemoglobin but refused further investigations. She continues to do well despite incidental low oxygen saturations on pulse oximetry.

(Figure 1.)



Discussion

A low oxygen saturation reading typically floods the mind of the physician with large list of possibilities. The list can be quite extensive, from external causes like movement artifacts and occluding nail polish

to internal causes ranging from pulmonary to cardiovascular to infectious diseases [8,9].

In certain situations, however, the clinical presentation of the patient may not tally with the readings obtained by pulse oximetry. Thus the pulse oximeter though very useful, has its accuracy limited in certain circumstances, especially in cases where the clinical appearance of the patient does not correlate with the readings [10,11,12].

A pulse oximeter measures absorbance by using 2 diodes transmitting light of differing wavelengths across a tissue to measure the intensity of the wavelengths at the other side. The percentage of oxygen saturation of haemoglobin can then be derived using the known absorbance spectra of oxygenated and deoxygenated haemoglobin [3,4,5].

In presence of a haemoglobinopathy, the haemoglobin molecule is altered, often as a result of an amino acid substitution that changes the molecule's affinity for oxygen. As a result pulse oximetry alone cannot reliably and accurately measure oxygen saturation.

Table 1. Causes and mechanisms of unreliable SpO_2 readings[8].

1.Causes of intermittent drop-outs or inability to read SpO_2	♀ Poor perfusion due to a number of causes, *e.g.*, hypovolaemia, vasoconstriction, etc
2. Causes of falsely normal or elevated SpO_2	♀ Carbon monoxide poisoning ♀ Sickle cell anemia , vaso-occlusive crises (overestimation of O_2Hb and underestimation of PaO_2)
3. Causes of falsely low SpO_2	♀ Venous pulsations ♀ Excessive movement ♀ Intravenous pigmented dyes ♀ Inherited forms of abnormal haemoglobin ♀ Fingernail polish ♀ Severe anemia (with concomitant hypoxemia)
4. Causes of falsely low or high SpO_2	♀ Methaemoglobinemia ♀ Sulfhaemoglobinemia ♀ Poor probe positioning ♀ Sepsis and septic shock
5. Causes of falsely low O_2Hb as measured by a co-oximeter	♀ Severe hyperbilirubinemia ♀ Fetal Hb (HbF)

In the case reported above, the patient was found to have Alpha variant haemoglobinopathy. With approximately 7% of the worldwide population being carriers, haemoglobinopathies are the most common monogenic diseases and one of the world's major health problems [13].

Defects in genes that control the expression of the haemoglobin protein can produce abnormal haemoglobins and anemia, which are conditions termed haemoglobinopathies. It includes all genetic haemoglobin disorders [14,15]. These are divided into two main groups as follows:

- Thalassemia syndromes
- Structural haemoglobin variants (abnormal haemoglobins)

Both are caused by mutations and/or deletions in the α- or β-globin genes [16,17]. When gene defects cause Hb synthesis disorders, this gives rise to thalassemias. Haemoglobin structure in these cases is normal. When they cause changes in Hb structure, it gives rise to abnormal haemoglobins [18,19]. There are also many mixed forms that combine features of both groups, e.g. β^0/β^+-thalassemias, HbSC disease and HbE α-thalassemias.

The clinical manifestations of the haemoglobinopathies are highly variable and range from mild hypochromic anaemia to moderate hematological disease to severe, lifelong, transfusion- dependent anaemia with multiorgan involvement [20].

Typically, structural haemoglobin variants are an autosomal dominant inherited haemoglobin disorder caused by structural defects resulting from an altered amino acid sequence in the α or β chains. Main types of abnormal haemoglobins are HbS (Sickle cell disease), HbE and HbC. Within these main types, there are several subtypes with differing disease patterns [20,21].

There are hundreds of haemoglobin variants that have been identified to date. While relatively rare, knowledge of haemoglobinopathies is important [22]. Some haemoglobin variants may cause haemolytic anemia while others cause methaemoglobinemia, cyanosis or polycythemia. In addition, there are those with a decreased oxygen affinity that often show no clinical symptomatology [21,22].

Many of these patients are asymptomatic and present with incidental findings of low oxygen saturations via pulse oximeter, but shows normal partial pressure of oxygen in the arterial blood. These patients tend to do quite well and are not truly hypoxic.

Though the use of pulse oximetry has greatly advanced our capabilities in vital monitoring, it's important to recognize its limitations. When asymptomatic patients present with low SpO2 readings via pulse oximetry and no clinical evidence to support the finding, the PaO2 should be measured and possibility of a haemoglobinopathy as a potential etiology is to be considered. Knowledge of these diagnoses can potentially spare patients unnecessary medical interventions and prolonged hospitalizations while providing the physician some ease of mind [22,23].

There is inadequate knowledge regarding the principles of pulse oximetry and also insufficient data on haemoglobinopathies in our setting [24,25,26]. Further, the optimal management of patients with abnormal haemoglobins remains to be defined [27,28,29]. Adequate studies to define and collate data on haemoglobinopathies in the Indian population can provide a unique opportunity to study the contributory effects of chronic hypoxia and haemolysis in cardio-pulmonary disease [30]. Better understanding and future research in this area can improve outcomes for the general population and especially for haemoglobinopathy patients.

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