CASE REPORTS

Anesthesiology
80:472–474, 1994

Unexpectedly Low Pulse Oximeter Readings in a Boy with Unstable Hemoglobin Köln

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PULSE oximetry is used widely to monitor hemoglobin oxygen saturation (Sp_{O_2}). We recently cared for a boy with hemoglobin Köln in whom Sp_{O_2} was unexpectedly low despite stable vital signs during surgery.

Case Report

A 9-yr-old boy (height 130 cm, weight 27 kg) with congenital microtia was admitted for reconstruction of the right ear. He had been in good health except for a history of occasional upper respiratory infection associated with occasional expiratory wheezes. A review of his medical records revealed that, soon after birth, he had been diagnosed with hemoglobin Köln inherited from his mother. In 1990, plastic surgery was scheduled to implant some costal cartilage in the ear. Preoperative laboratory findings were near normal except for total cholesterol (100 mg/dl), total bilirubin (1.8 mg/dl), and lactic dehydrogenase (301 mU/ml). The patient received intramuscular injections of scopolamine hydrobromide (0.25 mg) and hydroxyzine (25 mg) 1 h before surgery. Intermittent oscillometric blood pressure and rectal temperature were monitored in addition to electrocardiogram, and a pulse oximeter (Ohmeda Biox 3740, Louisville, CO) probe was attached. The Sp_{O_2} while breathing air upon entering the operating room was 89%. However, this was thought to be a false reading because of a poor fit of the sensor (Finger Clip 0380-1000-042, Ohmeda, Louisville, CO). Anesthesia was induced using intravenous diazepam (5 mg), followed by inhalation of nitrous oxide, oxygen, and halothane. Tracheal intubation was facilitated with an intravenous injection of succinylcholine chloride (30 mg). After intubation, Sp_{O_2} remained low, 90%, even at $F_{I_{O_2}} = 1$. Breath sounds were normal bilaterally, and the pulse oximeter displayed a two-dimensional plethysmographic waveform that appeared to be synchronous with the peripheral pulse. Before starting the operation, arterial blood was drawn at a fraction of inspired oxygen ($F_{I_{O_2}}$) of 0.5 and analyzed with a blood gas analyzer (NOVA STAT Profile 5, Boston, MA). The arterial oxygen tension (Pa_{O_2}) of 291 mmHg was considered sufficient for surgery, which was carried out uneventfully (table 1). Arterial blood at $F_{I_{O_2}} = 0.5$ was analyzed with a co-oximeter (Corning M 2500, Medfield, MA) soon after the appearance of spontaneous breathing, before the patient was awakened from anesthesia. Analytic data included: *p*H 7.323, Pa_{O_2} 206 mmHg, arterial carbon dioxide tension 48 mmHg, arterial oxygen saturation (Sa_{O_2}) 92.2%, carboxyhemoglobin (COHb) 3.8%, methemoglobin (MetHb) 1.0%, and reduced hemoglobin (HHb) 3.1%. The patient returned to the ward in good condition and recovered well except for mild anemia that probably was due to blood loss and hemolysis. He was discharged on the 30th postoperative day. These abnormal findings prompted us to study the patient and his mother the following year. Table 1 shows the patient's data both during surgery and later, and table 2 contains data from his mother. The mother's blood, drawn while she was breathing oxygen, also was analyzed in a second laboratory oximeter (Radiometer OSM_3, Copenhagen, Denmark). Findings from both were similar, with a slight increase in Sa_{O_2} as $F_{I_{O_2}}$ increased. Apparent COHb levels were about 4%, regardless of the $F_{I_{O_2}}$, except just after the operation, when it was 5.3%. However, none of his family had a history of smoking. In our case, abnormal hemoglobin content was 5.4% measured by

Table 1. Sp_{O_2}, Blood Gas Analyses, and Hemoglobin Fractions in the Patient with Hemoglobin Köln

	Beginning of OP	End of OP	Day after OP	1 yr Later at OPD	
$F_{I_{O_2}}$	0.5	0.5	Air	Air	1.0
Sp_{O_2} (%)*	≈89–90	90		87	92
*p*H	7.349†	7.323	7.451	7.482	7.521
Pa_{O_2} (mmHg)	291†	206	78	105	584
Pa_{CO_2} (mmHg)	43†	48	40	38	34
HCO_3 (mM)	23.9†	24.7	28.1	28.7	27.4
O_2Hb (%)		92.2	90.4	92.6	93.9
Sa_{O_2} (%)	99.9†	96.8	95.7	96.1	97.6
COHb (%)		3.8	5.3	4.0	3.8
MetHb (%)		1.0	0.2	0.1	0.0
HHb (%)		3.1	4.0	3.4	2.4
THb ($g \cdot dl^{-1}$)	12.8†	10.9	9.4	10.6	10.8

Measurements except as noted below were made with the Corning Co-oximeter M2500 (Medfield, MA). The values of Sa_{O_2} are calculated as $O_2Hb/(100 - COHb - MetHb)$.

OP = operation; OPD = outpatient department; $F_{I_{O_2}}$ = fraction of inspired O_2; Sp_{O_2} = O_2 saturation measured by pulse oximetry; Pa_{O_2} = arterial O_2 tension; Pa_{CO_2} = arterial CO_2 tension; O_2Hb = oxyhemoglobin; Sa_{O_2} = arterial O_2 saturation; COHb = carboxyhemoglobin; MetHb = methemoglobin; HHb = reduced hemoglobin; THb = total hemoglobin.

* Ohmeda 3740 with Finger Clip (0380-1000-042), Louisville, CO.

† NOVA STAT Profile 5, Boston, MA.

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Received from the Department of Anesthesiology, Fujita Health University School of Medicine, Aichi, Japan. Accepted for publication August 10, 1993. No grant or other financial support was received for this work. Presented at the annual meeting of the Japan Society of Anesthesiology, Tokai Branch, Toyoake, Aichi, Japan, February 16, 1991.

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Key words: Measurement techniques: pulse oximetry. Absorbance curves: hemoglobin Köln.

Table 2. Sp_{O_2}, Blood Gas Analyses, and Hemoglobin Fractions in the Patients' Mother with Hemoglobin Köln, Compared to a Nonsmoker with Hemoglobin A

	Hemoglobin Köln			Hemoglobin A	
$F_{I_{O_2}}$	Air	Air	1.0	Air	1.0
Sp_{O_2} (%)*	88	88	91	98	100
*p*H	7.441		7.513	7.431	
Pa_{O_2} (mmHg)	111.8		545.3	99.8	
Pa_{CO_2} (mmHg)	40.5		32.8	46.8	
HCO_3 (mM)	27.5		26.3	31.1	
O_2Hb (%)	92.8	93.7†	94.2	96.1†	98.1†
Sa_{O_2} (%)	96.9	97.8†	98.2	97.6†	99.9†
COHb (%)	4.2	3.7†	4.1	1.2†	1.3†
MetHb (%)	0.0	0.5†	0.0	0.4†	0.5†
HHb (%)	3.2	2.1†	2.0	2.3†	0.1†
THb ($g \cdot dl^{-1}$)	10.0	10.0†	10.2	14.6†	13.6†

Measurements except as noted below were made with the Corning Co-oximeter M2500 (Medfield,MA). The values of Sa_{O_2} are calculated as $O_2Hb/(100 - COHb - MetHb)$.

$F_{I_{O_2}}$ = fraction of inspired O_2; Sp_{O_2} = O_2 saturation measured by pulse oximetry; Pa_{O_2} = arterial O_2 tension; Pa_{CO_2} = arterial CO_2 tension; O_2Hb = oxyhemoglobin; Sa_{O_2} = arterial O_2 saturation; COHb = carboxyhemoglobin; MetHb = methemoglobin; HHb = reduced hemoglobin; THb = total hemoglobin.

* Ohmeda Biox 3740, with Finger Clip (0380-1000-042), Louisville, CO.

† Radiometer OSM_3, Copenhagen, Denmark.

the heat denaturation test.[1,2] Figure 1 shows the absorbance spectra for the hemoglobin fractions (O_2Hb, HHB, COHb) in the patient and a control. Samples from the patient and the control of hemoglobin A were diluted to 1.06 g/dl total hemoglobin content and further adjusted to *p*H 7.4 using TRIS buffer. Absorbance spectra were displayed by a visible recording spectrophotometer (UV 240, Shimadzu, Japan). All the curves for O_2Hb, HHb, and COHb were shifted upward in the patient with hemoglobin Köln compared with those for the control, ranging from 600 to 900 nm.

Discussion

Hemoglobin Köln constitutes about 10% of unstable hemoglobinopathies.[3] Patients with hemoglobin Köln present various degrees of abnormal hemoglobin content.[4] Laboratory oximeters such as the Corning M2500 and the Radiometer OSM_3 use multiple wavelengths[5] to separately determine O_2Hb, HHb, COHb, and MetHb. To distinguish between these terms for clarity, all international standards organizations have now agreed on the definition: $Sa_{O_2} = 100(O_2Hb)/(O_2Hb + HHb) = 100(O_2HB)/(100 - COHb - MetHb)$.

Since the Corning M2500 oximeter reports O_2Hb% as Sa_{O_2}%, the data were converted to conform with new standards. With these corrections, there was no significant difference between the results from the two oximeters.

How could the presence of only 5.4% of an abnormal hemoglobin cause an 8–11% error in the pulse oximeter reading? Examination of the O_2Hb spectrum in figure 1 at 660-nM wavelength shows an absorbance difference between the patient and control of 0.15 with only 5.4% hemoglobin Köln, which computes to an optical density of 2.7 for pure hemoglobin Köln. As this is unlikely, especially because the differences in O_2Hb absorbance were found across the spectrum, it may be necessary to consider other causes for this discrepancy. It is possible that the measured amount of hemoglobin Köln is an underestimate, because that species is thought to be fragile in the *in vitro* analysis. It also is possible that the analysis does not detect all the abnormal hemoglobin present. Both oximeters de-

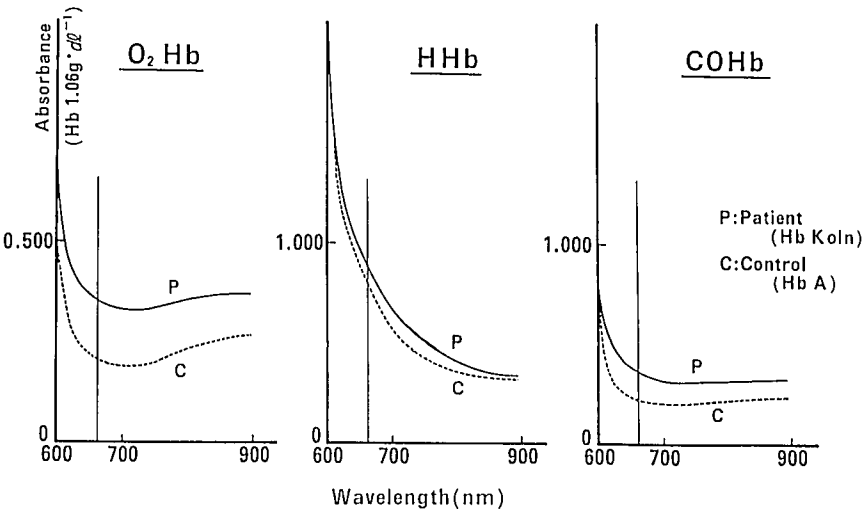

Fig. 1. The absorbance curves of hemoglobin fractions oxyhemoglobin (O_2Hb), reduced hemoglobin (HHb), and carboxyhemoglobin (COHb) in the patient with hemoglobin Köln, compared to a nonsmoker with hemoglobin A.

tect hemoglobin Köln primarily as if it were a mixture of COHb and HHb, somewhat more of the former.

The authors thank Professor Masami Hirano, Department of Hematology, Fujita Health University School of Medicine, for permission to present details of these hemoglobin Köln cases, and Professor Yashuhiro Shimada, Department of Anesthesiology, Nagoya University School of Medicine, for his advice to this study. Also, Dr. Itsuro Katsuta and his staffs at Fujita Health University School of Medicine College provided technical assistance in this investigation.

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Anesthesiology
80:474–476, 1994

An Unexpected Finding with Pulse Oximetry in a Patient with Hemoglobin Köln

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THE hemoglobinopathies characterized by an unstable hemoglobin, an array of more than 100 similar disorders caused by mutations of the hemoglobin molecule, are inherited in an autosomal dominant pattern and produce hemoglobins that denature spontaneously.[1] Clinically, these patients present with hemolytic anemia, which can be exacerbated by oxidants or infection.[1,2] However, their clinical course usually is benign.[1,2] An important feature of these disorders is variation in the affinity of the abnormal hemoglobin for oxygen.[1–6] An unexpected finding during the care of a patient with hemoglobin Köln, one of the hemoglobinopathies characterized by an unstable hemoglobin, was artifactually low hemoglobin oxygen saturation (Sp_{O_2}) recorded by pulse oximetry. It was hypothesized that methemoglobinemia produced by autoxidation of the unstable hemoglobin was responsible for the falsely decreased Sp_{O_2} observed.

Case Report

A 42-yr-old man presented for a unilateral Caldwell-Luc procedure for chronic sinusitis. At 26 yr of age, spherocytosis and splenomegaly had been found. Although asymptomatic, he underwent a splenectomy under an uneventful general anesthetic. Three years later, the patient's father had a myocardial infarction and was hospitalized. Routine complete blood count revealed polycythemia with a paradoxically elevated reticulocyte count of 5% in the father. A hematology consultant found the father's blood to have an increased oxygen affinity. Hemoglobin electrophoresis subsequently revealed an abnormal band characteristic of hemoglobin Köln, and additional questioning revealed that the patient's father was born in Germany. Subsequently, the hematology consultant performed hemoglobin electrophoresis on the father's immediate family, and the patient was found to have 30% hemoglobin Köln. Since the time of the patient's initial diagnosis, a daughter was born and found to have hemoglobin Köln.

The patient was otherwise healthy. He did not smoke cigarettes, specifically denied dyspnea on exertion or other pulmonary symptoms, and played tennis regularly. At the time of the Caldwell-Luc procedure, he had not seen a hematologist for more than 10 yr. He took no medications and denied any allergies. On physical examination, he was 196 cm tall and weighed 83 kg. Blood pressure was

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Received from the Department of Anesthesia, University of Pennsylvania Medical School, Philadelphia, Pennsylvania. Accepted for publication September 17, 1993.

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Key words: Blood, hemoglobin: hemoglobinopathy. Hemoglobin Köln. Monitoring: pulse oximetry.