Background:
Certain mutations occurring within the hemoglobin (Hb) can alter its structure and changes the stability of the molecule within the erythrocyte. This leads to the formation of an unstable compound which reduces erythrocyte deformability, increases splenic sequestration and causes hemolytic anemia. We report a case of a girl with Hb Southampton (also known as Hb Casper) in whom the pulse oximeter showed false low measurements.

Case:
A 5 year old Latino girl diagnosed with Hb Southampton presented for splenectomy due to numerous episodes of hemolytic anemia requiring monthly PRBC transfusions. The patient was transfused preoperatively and the surgical procedure was performed uneventfully. Postoperative Spo2 values ranged between 98%–100%.

Three months later the patient was diagnosed with pigment gallstone secondary to chronic hemolysis for which she underwent laparoscopic cholecystectomy with liver biopsy. After induction of general anesthesia via facemask, oxygen saturation (Spo2) values of 85-95% were noted. Adequate mask ventilation with Fio2 = 1 and change of probe site did not change the result (despite a good plethysmographic waveform). LMA was placed and secured with bilateral equal breath sounds. Arterial blood gas showed arterial oxygen saturation = 100% and PaO2 of 376 mmHg. The surgical procedure was then performed uneventfully. Postoperative Spo2 values ranged 87%–95%.

Discussion:
Continuous pulse oximetry is a widely accepted method of accurately estimating hemoglobin oxygen saturation. There have been a limited number of case reports of unstable Hb variants with unexpectedly low pulse oximetry values.

Hb Southampton is a rare variant that arises from a mutation characterized by the substitution of a leucine residue for a proline at codon β106 (CTG→CCG). This mutation breaks the G helix and severely distorts the tertiary structure of the molecule producing an unstable Hb. (1). The resultant unstable Hb molecule precipitates within the erythrocyte causing intracellular inclusions. Cells containing precipitates are either sequestered within the spleen, or destroyed, resulting in a hemolytic anemia.

In the first 5 years of life, monthly PRBC transfusions caused iatrogenically high concentrations of stable Hb A which affected the pulse oximetry values. Once splenectomy had been performed and the patient no longer required monthly transfusions, the unstable Hb variant was no longer sequestered by the spleen, and its predominance within the circulation led to low pulse oximetry values. It is possible that Hb Southampton is not absorbed at either of the two wavelengths, or that oxidation of the Hb variant secondary to infection or oxidizing medications causes formation of significant amounts of metHb.

In summary, a pulse oximeter is not an accurate monitor for patients with Hb Southampton and possibly other hemoglobinopathies. In the future, the presence of any abnormal Hb molecules should warrant a discussion about additional means for effectively and accurately monitoring oxygen content of blood.

References:
(1)Hemoglobin Southampton (Casper): characterization of the base mutation.